THE MEDIASTINUM

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To Our Wives

PREFACE

THIS MONOGRAPH is intended for all physicians interested in mediastinal lesions but particularly for those who are charged with the responsibility of accurately diagnosing and treating chest diseases. The authors recognize that the diagnosis of mediastinal lesions in many instances can not be made short of surgical exploration. Nevertheless many of the features of a lesion can often be recognized on radiologic examination. There is little doubt that radiologic procedures can many times determine the characteristics and anatomic relationships of mediastinal masses.

The mediastinum is a relatively small compartment of the body but contains a large number of important anatomic structures. Pathologic changes within or adjacent to the mediastinum may involve these structures primarily or secondarily. In this monograph we have included not only the common types but many of the rare lesions as well and have attempted to discuss these entities in an orderly and simplified manner including in these discussions both general considerations as well as radiologic characteristics.

One important organ of the mediastinum—the heart—has been deliberately excluded. Heart disease is a subject within itself and discussions of cardiac lesions do not belong in a text of this type. However mass lesions of the pericardium and vascular lesions of the mediastinum are discussed in as much as they frequently enter into the differential diagnosis of mediastinal disease. It is admitted that the selection of the subject matter had to be somewhat arbitrary.

We wish to give credit to the following individuals who have been of so much help in the preparation of this monograph. For secretarial work: Miss Ethyl Fitzpatrick and Miss Sue Hodges. For some of the drawings, sketches, photographs and other medical art work: Miss Kathleen Mackay and Mr Joseph Jackson. For critical analyses of certain of the material in the text: Doctors Charles M Huguley Jr, James V Rogers Jr, Brit B Gay Jr, Osler A Abbott and William E Van Fleet.

We are also indebted to many authors of articles published in recent years who allowed us to use illustrations from their papers and to the editors of various medical publications who granted us permission to reproduce this material. Our thanks also go to many medical friends who were kind enough to furnish us the roentgenograms of unpublished cases. For all of these proper credit is given in the legends under the illustrations.

CONTENTS

THE MEDIASTINUM

Chapter I

THE ANATOMY OF THE MEDIASTINUM

THE MEDIASTINUM is the subdivision of the thorax which is located between the pleural cavities. It extends from the sternum anteriorly to the vertebral column posteriorly. The upper limit of the mediastinum is formed by the thoracic inlet, whereas the lower border plane of the mediastinum is represented by the diaphragm.

This chapter gives a brief description of the gross anatomic and principal radiologic features of the mediastinum. The anatomy of the mediastinal lymph nodes is discussed in a separate chapter because of the importance of mediastinal lymphadenopathy in pulmonary diseases.

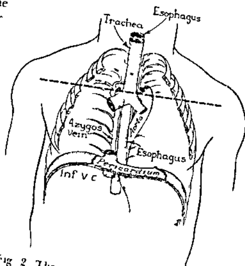


Fig 2 The upper and lower compartments of the mediastinum in the frontal projection

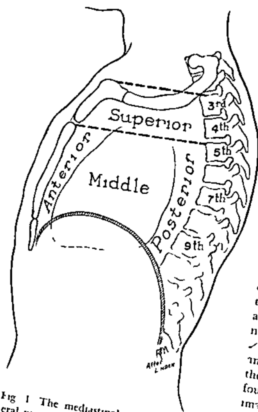
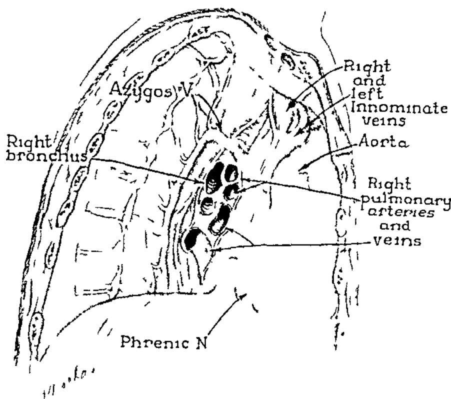


Fig 1 The mediastinal subdivisions in the lateral projection

Arbitrarily the mediastinum has been subdivided into several compartments. A plane which extends from the sternal angle to the fourth intervertebral disc subdivides the mediastinum into an upper and lower division. The upper division is designated as the superior mediastinum. The lower compartment is again subdivided into three spaces which are described as the anterior, middle and posterior mediastinum.

The *superior mediastinum* is bounded anteriorly by the sternum, superiorly by the thoracic aperture, posteriorly by the upper four dorsal vertebrae and inferiorly by an imaginary plane extending from the sternal angle to the fourth intervertebral disc. Anteriorly behind the sternum are found the thymus or its remnants, the innominate



A

Fig 3 The right lateral aspect of the mediastinum as seen in the right lateral and rigl anterior oblique view

veins and superior vena cava The aortic arch and its major branches occupy a slightly more posterior position in front of the trachea The vagi the left recurrent laryngeal nerve and the phrenic nerve are situated in close relationship to the large vessels Other vascular structures of the superior mediastinum are the arch of the azygos vein and the upper segment of the hemiazygos system The space between trachea and spine is occupied by the esophagus and thoracic duct

The lymph nodes of the superior mediastinum are found to be adjacent to the large vessels the trachea and spine The internal mammary vessels and lymph nodes are placed slightly lateral to the superior mediastinum though they are frequently in

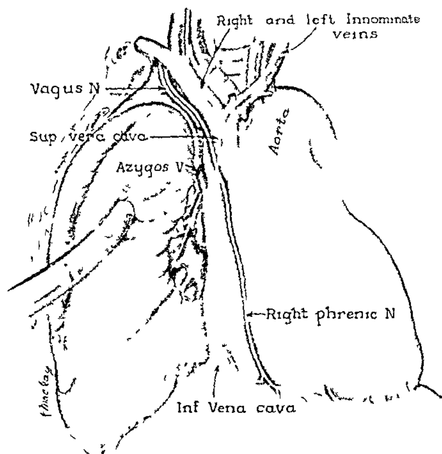


Fig 3B

cluded in a description of the superior mediastinal structures The same applies to the upper segments of the sympathetic chains which are likewise situated lateral to the superior mediastinum behind the parietal pleura

The *anterior mediastinum* is a shallow space which is bounded anteriorly by the sternum and the transverse thoracic muscle and posteriorly by the pericardium At the level of the second third and fourth costal cartilages this space is very narrow To wards the diaphragm the space becomes wider as the left pleural sac recedes laterally to accommodate the pericardium

The contents of the anterior mediastinum are the lower portion of the thymus gland or its remnants a few lymph nodes anterior to the pericardium the sterno pericardial ligaments and the left internal mammary vessels and lymph nodes The right internal mammary vessels and lymph

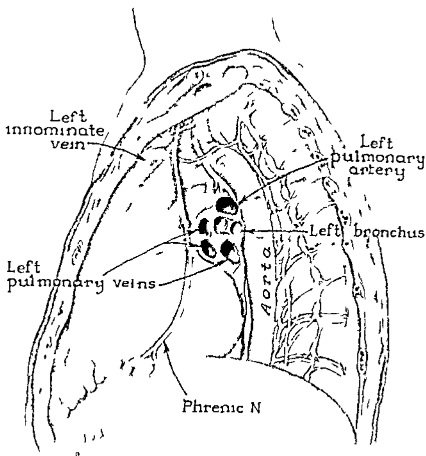


Fig 4 The left lateral aspect of the mediastinum as seen in the left lateral and left anterior oblique views

nodes cannot be considered as part of the *anterior mediastinum* since they are bounded posteriorly by the pleural sac and thoracic muscles

The *middle mediastinum* is largely formed by the pericardial sac and tracheal bifurcation The anterior and posterior portions of the pericardial sac represent therefore the anterior and posterior borders of this mediastinal space Its superior and inferior borders are the same as that of the *anterior mediastinum* The contents of the middle mediastinum are all structures enclosed in the pericardium the roots of the lungs the pulmonary arteries and veins the phrenic nerves and tracheobronchial lymph nodes

The *posterior mediastinum* extends from the posterior aspect of the pericardium to the spine Its superior and inferior borders

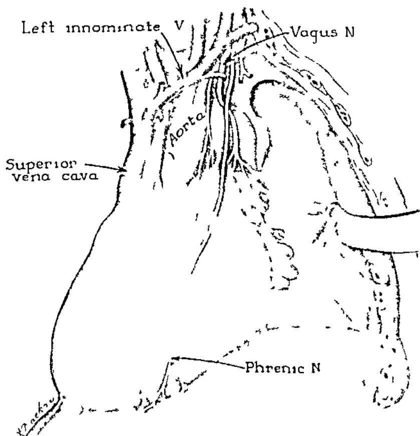


Fig 4B

correspond to those of the anterior and middle mediastinum

The posterior mediastinal contents are the lower esophagus and the descending aorta Other vascular structures are the intercostal peribronchial pericardial esophageal and mediastinal arteries and their companion veins The posterior mediastinum contains also the vagi and the thoracic duct Posteriorly are found the sympathetic chains the paravertebral lymph nodes as well as the azygos and hemiazygos system

The anatomic classification of the mediastinum presented here is clearly an artificial one It should be noted that the superior mediastinum communicates widely with the other mediastinal partitions as well as the visceral compartments of the neck This lack of anatomic barriers enhances the spreading of infection and neoplastic disease throughout the mediastinal compartments and adjacent cervical spaces

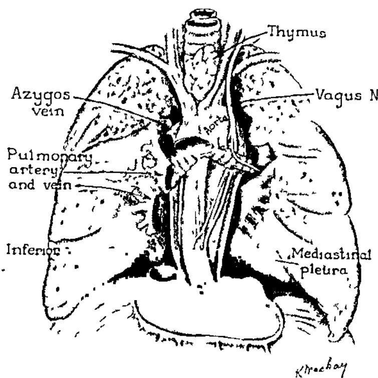


Fig 5 Anterior view of the posterior and superior mediastinum

For an understanding of the radiologic features of the mediastinum this space may be envisaged as an irregularly wedge shaped body which has a wide anterior and narrow posterior segment The lateral walls show several projections and indentations reflecting the shape of the mediastinal organs Not all of the curved planes of the lateral mediastinal borders are readily demonstrated on radiologic examinations Obviously those structures which protrude most prominently into the adjacent lung fields are radiologically most distinct Thus in the frontal roentgenograms the right border of the mediastinum is seemingly made up by the right innominate vein the superior vena cava and the right heart border, whereas the left margin of the mediastinum appears to be formed by the left subclavian vessels, the aorta, the pulmonary artery and the left cardiac border

A more detailed radiologic investigation of the mediastinum may be carried out with the aid of grid films, high kilovoltage techniques tomography and contrast visualization of the esophagus In this manner several mediastinal contour lines becomes visible within the mediastinal silhouette The

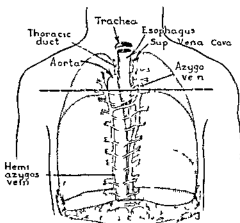


Fig 6 Posterior view of the posterior mediastinum

borders of these structures against the adjacent lung are often less conspicuous as they may be superimposed upon the shadows of the heart spine and large vessels

Roentgen studies of the various mediastinal contour lines have made many contributions to the anatomy of the mediastinum in the living and brought out certain differences with classic anatomic concepts based on cadaver observations At the same time the analysis of these mediastinal borders have proved to be clinically important and any unusual bulge or interruption of their continuity may indicate disease processes of the mediastinum or adjacent pleura or lung

Some of the more important radiologic features of the mediastinum shall be described in the following

Anatomic cross sections of the *superior mediastinum* frequently show the esophagus close to the spine with the lungs widely separated by intervening tissue Contrary to this concept roentgen studies during life



Fig 7 The contour lines of the posterior mediastinum superimposed upon the tracheal air shadow (arrows) This thin curvilinear shadow is caused by the normal extension of the upper lobes into the prevertebral space



Fig 8 Tomogram of superior mediastinum The prevertebral mediastinal space between the two upper lobes measures only a few millimeters in thickness and is indicated by arrows



Fig 9 The soft tissue space of the anterior mediastinum is outlined on tomography (arrows)



Fig 10 Appearance of the anterior mediastinum in a patient with emphysema The arrows indicate the septum like structure

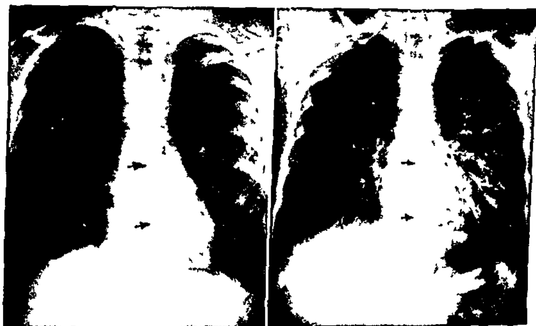


Fig 11 The right lateral aspect of the posterior mediastinum is clearly visible against the right lower lobe (arrows) The bronchogram shows the prevertebral position of the right lower lobe in this child Contrast medium in the esophagus shows that its right lateral wall forms the right lateral border of the posterior mediastinum in this region

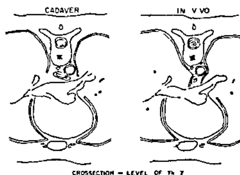


Fig 12 Cross section of the mediastinum at the level of the seventh dorsal vertebra Many anatomy texts depict the space between the heart and spine (posterior mediastinum) as a broad tissue structure separating the lungs widely During life the posterior mediastinum is a narrow space with varying degrees of pulmonary incursion into the prevertebral region

indicate that the esophagus is sufficiently anterior to the dorsal spine to allow both lungs to approximate each other in the prevertebral region The medial borders of both lungs may then be superimposed upon the tracheal air shadow The space between the lungs may measure from a few millimeters to very few centimeters in width This may be shown on conventional roentgenograms Tomograms reveal readily that the superior mediastinum behind the esophagus is a rather narrow mesentery like structure during life

The very shallow *anterior mediastinum* is visualized only with great difficulty This potential space may be demonstrated to advantage on roentgenograms taken in lateral projections on deep inspiration If the patient assumes the recumbent position a further separation of the structures of the middle mediastinum from the sternum may occur and aid in the demonstration of retrosternal masses

In infancy and early childhood the thymus gland may extend into the anterior mediastinum causing a distinct shadow on roentgenograms both in the frontal and lateral projections In the adult this tissue space contains only remnants of the thymus and alveolar tissues and may be demon

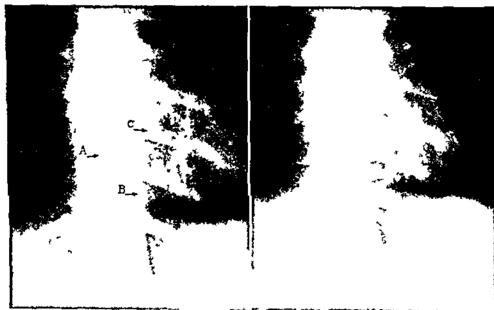


Fig 13 The contour lines of the posterior mediastinum A The border of the mediastinum against the right lung B The paraspinal soft tissue line on the left side C The left lateral wall of the aorta The right lateral wall of the aorta is not visible and the narrow space between lines A and C does not reflect the aortic width

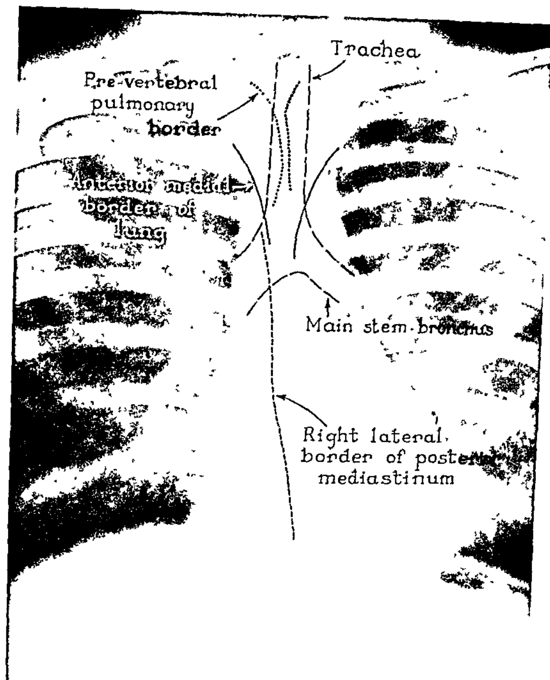


Fig 14 A schematic outline of the contour lines of the mediastinum

strated in frontal roentgenograms by the *antero-medial borders of the lungs* In patients with emphysema the upper lobes may be in contact in the region of the retrosternal space and part of the anterior mediastinum may appear as a thin septum These mediastinal contour lines which are particularly well shown on tomography may intersect with the contour lines of the *posterior mediastinum*

A detailed discussion of the *middle mediastinum* containing largely the heart and pericardium would go beyond the scope of this book It should be emphasized here that the right lateral contour of the left atrium as well as a segment of the large

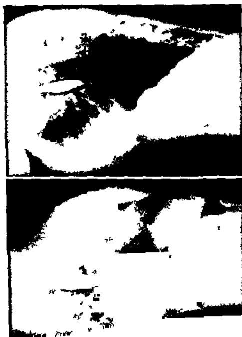


Fig 15 Esophagogram in prone and supine position indicating considerable flexibility of the esophagus and posterior mediastinum The mesentery like character of the posterior mediastinum allows a close approximation of both lungs in this region

veins entering the atrium is visible in a small percentage of normal individuals Demonstration of a double contour of the right heart border is therefore not necessarily indicative of mitral valvular disease or left atrial enlargement

The *posterior mediastinum* extends from the posterior aspect of the pericardium to the spine Its superior and inferior borders correspond to those of the anterior and middle mediastinum

The posterior mediastinal contents are the lower esophagus and the descending aorta Other vascular structures are the intercostal, peribronchial pericardial esophageal and mediastinal arteries and their companion veins The posterior mediastinum contains also the vagi and the thoracic duct Close to the spinal column are found the sympathetic chains the paravertebral lymph nodes as well as the azygos and hemiazygos system

The roentgen findings also indicate that the posterior mediastinum is not located in midline but somewhat towards the left side At the same time it is slightly tilted Contrary to cadaver observations the width of the posterior mediastinum may not measure more than 2 cm



Fig 16 The inner and outer surface of the right tracheal wall is clearly visible (arrows) With the development of large paratracheal lymph nodes the outer contour of the trachea has disappeared The right lung is no longer in contact with the right tracheal wall



Fig 17 Right and left paraspinal soft tissue lines (arrows)

Contrast examination shows the *esophagus* to be cylindrical with the exception of studies during life indicate that the esophagus is suspended in the superior and posterior *mediastinum at some distance from* the spinal column Roentgenograms taken in recumbent and prone position reveal considerable flexibility of this structure within the mediastinum

Contrast examination shows the esophagus to be cylindrical with the exception of those segments which show shallow indentations and deviations by adjacent organs The right wall of the esophagus may be visible against the lung in front of the spine The left wall of the esophagus may be outlined in the upper thoracic region above the level of the aortic arch In its upper portion the esophagus is not strictly in midline beyond the trachea but slightly placed to the left side

The following indentations and deviations of the esophagus may be normally observed in the mediastinum

1 A left concave indentation by the aortic arch

2 An anterior concave indentation by the left main bronchus close to the bifurcation of the trachea

*3 A shallow indentation which is an*teriorly concave by the posterior aspect of the heart notably the left atrium

4 A posterior concavity caused by the aorta This indentation varies considerably depending upon length and width of the descending aorta

The thoracic segment of the *trachea* reveals a distinct indentation on the left side caused by the adjacent aortic arch Opposite to this indentation a shallow impression by the azygos vein occasionally may be noted Generally the air column of the trachea permits a distinct visualization of the position and diameter of this structure In some instances however mediastinal contour lines may be superimposed upon the trachea simulating narrowing or unusual deviations They are formed by extension of the lungs into the retrosternal and retroesophageal spaces of the mediastinum

On the right side both the inner and outer walls of the trachea are usually demonstrated In this region the trachea with the overlying mediastinal pleura is in direct *contact with the right lung* On the left side blood vessels are placed into the space between the trachea and left lung so that the outer wall of the trachea is usually not depicted If—in a series of roentgenograms—the outer wall of the trachea on the right becomes indistinct or obliterated enlargement of vascular structures enlargement of lymph nodes or development of other soft tissue masses should be suspected

On the left side of the spine from the level of the aortic arch to the diaphragm a sharply defined soft tissue band is frequently observed on the frontal roentgenograms of the chest and spine A similar shadow is less frequently demonstrated on the right side Most authors agree that these

shadows are cast by the soft tissues beneath the parietal pleurae covering the spine

On the right the parietal pleura covering the posterior chest wall and lateral spinal column proceeds in an oblique plane towards the midline before it reflects sharply to the mediastinal pleura On the left side however the vertebral pleura blends gradually with the mediastinal pleura maintaining a much more sagittal direction This more sagittal arrangement of the pleura on the left side enhances its roentgen demonstration by the tangential x ray beam and may explain why this shadow is seen more frequently on the left than on the right side

In individuals with a tortuous aorta the left paraspinal shadow is very often widened This is apparently caused by the tendency of the aorta to dislodge the pleura away from the spine Clinical observations of the paraspinal shadows carry considerable significance as localized or diffuse bulges of these shadows may indicate pathologic processes of the posterior mediastinal or spinal space On the other hand a sharp interruption or obliteration of the paraspinal shadow may denote loss of air content in the adjacent pulmonary tissue or pleural fluid formation

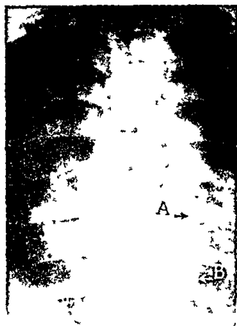


Fig 18 The left paraspinal line is often found at some distance from the spine in patients with tortuosity of the aorta A Paraspinal line B Left lateral wall of descending aorta

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Chapter 2

THE ANATOMY OF THE MEDIASTINUM (CONTINUED) THE MEDIASTINAL LYMPH NODES

THE LYMPH NODES of the thorax form an extrathoracic and intrathoracic group Only those chains of intrathoracic nodes which have a close relationship to the mediastinum shall be considered here

A THE PARIETAL LYMPH NODES

The parietal lymph nodes may be subdivided into the following chains

1 The anterior parietal or internal mammary lymph nodes
2 The posterior parietal nodes
3 The diaphragmatic lymph nodes

1 The anterior parietal or internal mammary nodes have attracted considerable interest on account of their involvement in neoplastic diseases of the breast Actually most of these nodes are not within the confines of the mediastinum being located behind the intercostal spaces and costal cartilages in front of the pleural spaces laterally or medially to the internal mammary vessels The internal mammary nodes are found with considerable regularity at the level of the first and second interspaces Their presence in the third to sixth interspace is less consistent The upper nodes are usually somewhat larger than the lower ones In addition to the internal mammary chain a few lymph nodes may be found directly behind the sternum within the anterior mediastinal space

These lymph nodes receive lymphatics

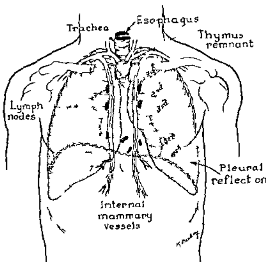


Fig 19 The lymph nodes of the anterior mediastinum The lymph nodes are found in close relationship to the internal mammary vessels



Fig 20 Metastatic involvement of the upper left internal mammary chain in a patient with carcinoma of the breast (arrows)

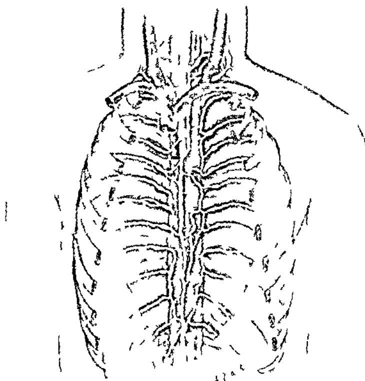


Fig 21 The thoracic duct and lymphatic channels of the posterior mediastinum The lymph nodes are in close relationship to the spine

from the anterior thoracic and abdominal walls and mammary glands Also by way of the diaphragmatic nodes the superior and inferior surface of the liver may drain into the internal mammary nodes

The terminal trunks of this lymph node system empty into the thoracic duct on the left and into the right lymphatic duct on the right side There are many variations with direct drainage of these channels into the jugulo subclavian angle or nodes of the cervical chains

2 The posterior parietal nodes consist of the intercostal nodes and juxta vertebral nodes Only the latter ones may be contained in the mediastinum The lymph nodes anastomose freely and receive lymphatics from the intercostal spaces pleura vertebrae and spinal muscles The efferent lymph vessels drain directly or indirectly

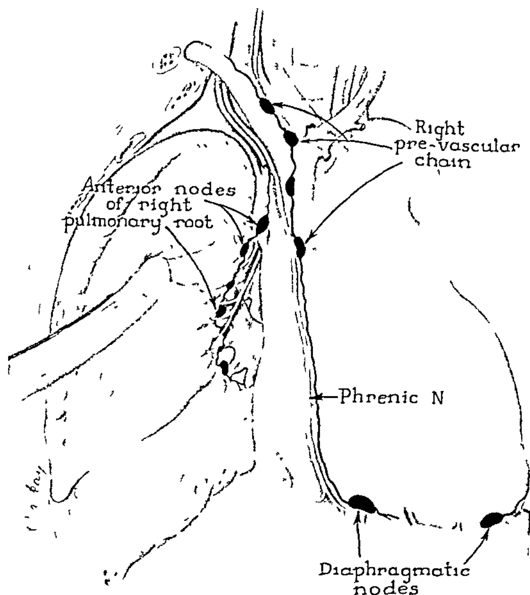


Fig 22 The prevascular and diaphragmatic lymph nodes on the right side The position of the anterior nodes of the right pulmonary root is also indicated

into the thoracic duct In the upper spinal region there are usually anastomoses to lymph trunks emptying into the right jugulo subclavian angle or into the lymph node chains of the neck

3 The diaphragmatic lymph nodes are found along the diaphragmatic aspect of the mediastinum usually anteriorly and laterally to the attachment of the pericardial sac. The nodes receive lymph vessels from the diaphragm and its covering as well as from the subphrenic structures such as the liver The efferent vessels drain into the internal mammary node system or as-

cend with the phrenic nerves and vessels to the anterior mediastinum

B THE VISCERAL INTRATHORACIC LYMPH NODES

The visceral intrathoracic lymph nodes comprise the following groups

1 Anterior mediastinal or prevascular lymph nodes
2 Posterior mediastinal lymph nodes
3 Peritracheal bronchial lymph nodes
4 Intrapulmonary lymph nodes

1 The anterior mediastinal or prevascular lymph nodes form chains which are gen

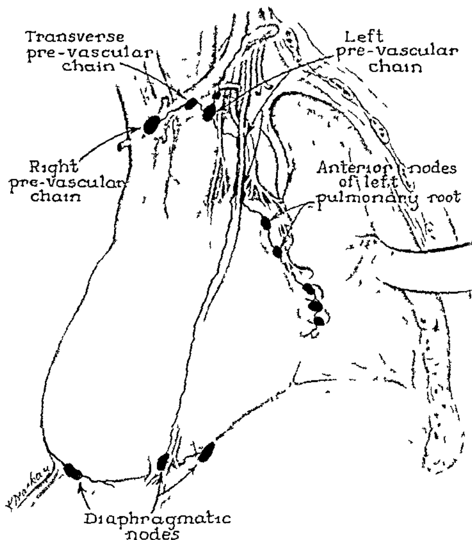


Fig 23 The left diaphragmatic and prevascular lymph node chain



Fig 24 The position of the right prevascular chain is indicated by silver clips placed at the anterior aspect of the superior vena cava and right innominate vein (arrows)

erally located along the anterior aspect of the large vessels On the right side this group of lymph nodes has a close relationship to the superior vena cava right innominate vein and innominate angle

On the left side the lymph nodes are found to be preaortic and precarotid Another chain is arranged in a transverse manner behind the thymus gland linking the two prevascular groups

These nodes receive afferent vessels originating in the anterior parts of the pul



Fig 25 A group of small calcified lymph nodes of the right prevascular chain Note that these nodes are at some distance from the trachea in the lateral view (arrows)

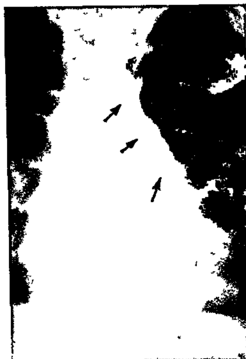


Fig 26 Calcified lymph nodes of the left prevascular chain (arrows) in a child with pulmonary tuberculosis



Fig 27 A large lymph node in the region of the left prevascular group This node is most likely located in the region of the ligamentum arteriosum (Botalli s node)

monary roots diaphragmatic and mediastinal pleurae heart, pericardium thymus the thyroid gland and trachea The efferent vessels terminate usually in the thoracic duct or directly into the jugulo-subclavian angles Occasionally drainage into the jugulo lymph node chains is found

2 The posterior mediastinal lymph nodes are located adjacent to the lower esophagus and descending aorta being more numerous in the lower portion of the mediastinum The afferent vessels of these nodes issue from the diaphragm the esophagus pericardium and lower lobes of the lungs The efferent channels empty into the nodes of the tracheal bifurcation or directly into the thoracic duct

3 The peritracheal bronchial nodes are subdivided into the following groups

(a) The paratracheal nodes

(b) The nodes of the bifurcation

(c) The nodes of the pulmonary roots

(a) The paratracheal nodes are found on both sides behind the large vessels close to the trachea and recurrent laryngeal nerve The right chain is usually more developed Its lowest node which may be fairly large is situated adjacent to the arch of the azygos vein and commonly referred to as the azygos node Less consistently a chain of small nodes may be found directly behind the trachea

The right nodes form an ascending lymphatic pathway which receives vessels directly from the right lung or from the nodes of the pulmonary root and bifurcation The lymphatics of the trachea esophagus and thymus also enter these lymph nodes Generally the efferent vessels form trunks which drain with many variations into the right jugulo subclavian venous system

The left paratracheal nodes form a similar ascending pathway which empties usually into the thoracic duct and w

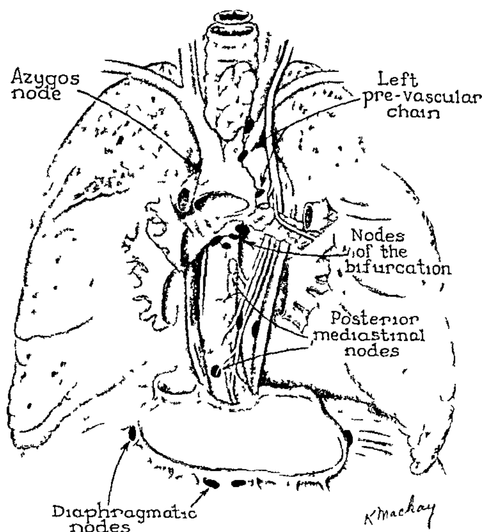


Fig 28 The lymph nodes of the posterior mediastinum

ceives lymphatics from the trachea the esophagus and left lung

(b) The nodes of the tracheal bifurcation consist of one rather large and several small nodes along the inferior and anterior aspect of the tracheal bifurcation These nodes accept lymphatics which stem from the diaphragm heart pericardium esophagus and both lungs The efferent lymph channels terminate usually in the right paratracheal chain

(c) The nodes of the pulmonary roots surround the hilar structures in close relationship to bronchi arteries and veins As a rule an anterior posterior superior and inferior group is distinguished Some of

the inferior nodes are placed in the pulmonary ligaments where it may be difficult to separate them from the posterior mediastinal lymph node group The nodes of the pulmonary roots are considered to be within the confines of the mediastinum They receive lymphatics from the pulmonary lobes and drain into the paratracheal nodes and nodes of the bifurcation

4 The intrapulmonary lymph nodes consist of the intrapulmonary nodes proper and the superficial subpleural nodes which will not be considered here

The intrapulmonary nodes are usually situated in the angles of the bronchial arterial and venous divisions They may be localized in between the lobes (interlobar) or within the lobes (lobar) On the right side a superior interlobar node is placed in the angle between the upper lobe bronchus and mainstem bronchus A right inferior interlobar node is found just below the origin of the middle bronchus

On the left side an interlobar node occurs in the angle formed by the bronchi of the upper and lower lobes

Other interlobar nodes are developed at



Fig 29 Calcified posterior mediastinal lymph nodes (arrows) The lymph nodes are in close relationship to the esophagus which is faintly opacified with contrast medium



Fig 30 Marked enlargement of posterior mediastinal lymph nodes in a patient with lymphoma Note the pressure defect on the esophagus



Fig 31 A small para-aortic node (arrows) in a patient with pulmonary carcinoma

the subdivisions of the bronchi and arteries located either in the pleural fissures or pulmonary parenchyma

Actually the interpulmonary nodes constitute lymphoid tissue outside the mediastinal space They represent however the primary drainage area of the lymphatics of the lungs and are usually in direct continuity with the mediastinal lymph nodes of the pulmonary roots

C THE LYMPHATIC DRAINAGE OF THE LUNGS AND ESOPHAGUS

In neoplastic and inflammatory diseases of the lungs and esophagus involvement of the regional lymph nodes is of considerable diagnostic and therapeutic importance The normal lymphatic drainage of the organs shall be briefly summarized Admitted there are many anatomic variations Also with regard to the intrapulmonary lymphatic drainage appreciable differences of normal anatomic concepts exist

Under pathologic conditions blockage of lymph nodes may enhance collateral and retrograde flow of lymph which in itself may explain deviations from predicted patterns in the lymph node spread of pulmonary cancer

According to Rouvière each lung consists of three principal drainage regions a superior middle and inferior one which do not correspond to the lobar subdivisions Lymph passes from these regions directly or indirectly by way of nodes within the lung or pulmonary roots to regional drainage chains in the mediastinum

From the right superior region (antero-

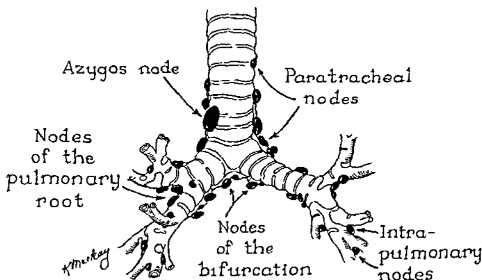


Fig 32 The peritracheal bronchial lymph nodes



Fig 33 Enlargement of right paratracheal nodes (arrows) There is also marked hilar adenopathy present



Fig 34 Enlargement of the bifurcation and posterior mediastinal nodes The esophagus is displaced posteriorly

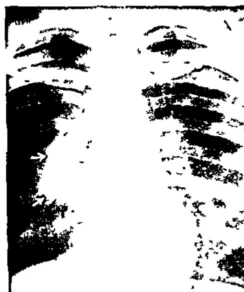




Fig 35 Enlargement of the nodes of the pulmonary root and of the intrapulmonary nodes in a patient with sarcoid (arrows)

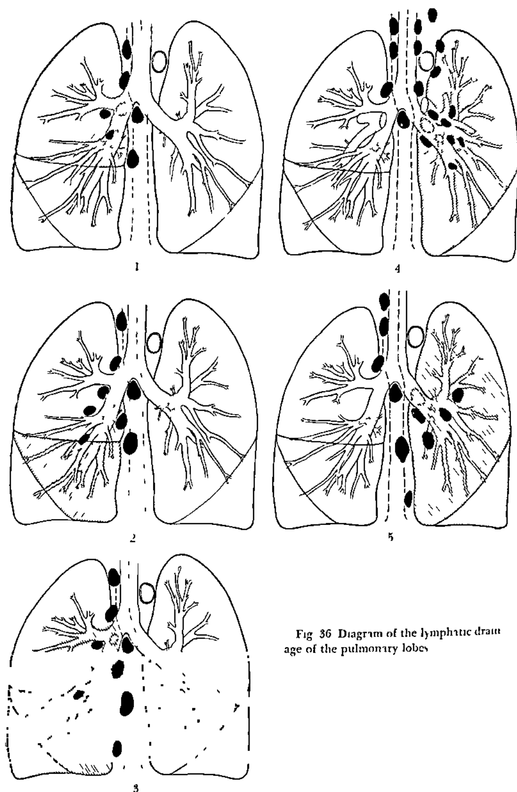


Fig 36 Diagram of the lymphatic drainage of the pulmonary lobes

medial aspect of the right upper lobe) lymph flows to the right paratracheal lymph node group The middle region (postero lateral aspect of the right upper lobe the right middle lobe and upper part of the right lower lobe) drains lymph into the right paratracheal chain and nodes of the bifurcation The inferior region (base of the right lower lobe) empties lymph into the nodes of the bifurcation and posterior mediastinal nodes adjacent to the esophagus

On the left side lymph passes from the superior region (upper portion of the left upper lobe) to the left prevascular and paratracheal lymph node groups The middle region (lower part of the left upper lobe and superior and middle zones of the left lower lobe) has the left paratracheal and bifurcation lymph nodes as drainage area The inferior region (basilar part of the left lower lobe) drains lymph into the bifurcation nodes and juxta esophageal nodes

With these observations Rouviere has shown that a major portion of the left lung will direct its lymphatic channels toward the bifurcation lymph nodes which in turn drain into the right paratracheal chain The right paratracheal chain represents therefore a principal drainage area for the entire right lung and a major portion of the left lung This may explain the frequent predominant involvement of right paratracheal lymph nodes in various types of intrathoracic diseases

McCort and Robbins and more recently Nohl have carefully analyzed the lymph node spread of pulmonary cancer Their observations tend to confirm the anatomic concepts presented by Rouviere

The thoracic esophagus has two extensive lymphatic networks the submucous and muscular lymphatic systems The lymphatic channels of these two systems traverse not only the entire length of the esophagus but also anastomose with each other Some of the collecting vessels of these networks may pass directly to the nearest regional nodes Others may ascend or descend for some distance before emptying into adjacent lymph node chains of the neck mediastinum or epigastrium

The echelon of lymph nodes which receive lymph from the esophagus are from above downward The internal jugular chains both paratracheal lymph node groups the nodes of the tracheal bifurcation the posterior nodes of the pulmonary roots the posterior mediastinal nodes the nodes of the cardia and lesser curvature of the stomach

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Chapter 3

MEDIASTINAL EMPHYSEMA

MEDIASTINAL emphysema or pneumomediastinum is the abnormal collection of air or gas in the tissue spaces of the mediastinum. Though mediastinal emphysema may exhibit a number of characteristic clinical symptoms and signs, its diagnosis usually depends on radiologic examination. In many cases pneumomediastinum may be observed as an incidental finding of little clinical importance. In other instances, however, mediastinal emphysema may predominate the clinical picture on account of its adverse effects upon the cardiovascular and respiratory systems. On rare occasions mediastinal emphysema is a highly significant finding as it may be an important clue to the diagnosis of rupture of a cervical, mediastinal or abdominal viscus. The radiologic recognition of mediastinal emphysema may also be exceedingly important in the differential diagnosis of other acute conditions such as coronary artery disease, acute pericarditis, pulmonary infarction and spontaneous pneumothorax.

The pathways of air into the mediastinum and the causes of mediastinal emphysema shall be considered together.

Air may find access to the mediastinum from the soft tissues and organs of the neck, the chest wall and pleural cavities, the subphrenic spaces, the mediastinal viscera and the interstitial spaces of the lung. Rarely bacterial gas formation will lead to the development of mediastinal emphysema in the course of mediastinitis or abscess.

The passage of air from the fascial com



Fig. 37. Development of extensive mediastinal emphysema and pneumothorax following tracheostomy.



Fig. 38. Very marked mediastinal emphysema following chest trauma. In the lateral view most mediastinal structures are outlined with unusual clarity.



Fig 39 Spontaneous rupture of the esophagus This 81 year old patient developed sudden upper abdominal and left anterior chest pain following a bout of pneumonia accompanying an epileptic seizure Note air in the mediastinum and soft tissues of the neck (arrows) There is a left pneumothorax and pleural effusion



Fig 40 Localized mediastinal emphysema A 20 year old male developed sudden severe anterior chest pain following diving into a swimming pool Faint crackling sounds were heard with each systole behind and to the left of the sternum Arrows indicate the mediastinal emphysema and displaced mediastinal septum

partments of the neck into the superior mediastinum is frequently observed particularly following surgical procedures of the head and neck Mediastinal emphysema is a common event in patients with tracheostomy It is less frequently encountered secondary to thyroidectomy radical neck dissection tonsillectomy and cervical neurosurgical procedures In these instances air dissects into the thorax passing between the pretracheal and prevertebral layers of the deep cervical fascia into the superior mediastinum A similar course of events takes place in patients with laceration of the cervical soft tissues or viscera

Injuries of the chest wall as well as thoracic surgical procedures may enhance the access of air into the mediastinum Mediastinal emphysema is not uncommonly noted in patients with rib fractures and pneumothorax Following dorsal sympathectomy air may be discovered in the mediastinum as an incidental finding

Pneumomediastinum is known to have occurred as the result of passage of air from the subphrenic space into the mediastinum Thus air in the mediastinum is not infrequently detected following injection of air or gases into the peritoneal cavity or retroperitoneal space Aside from these diagnostic and therapeutic procedures pneumomediastinum is occasionally discovered following perforation of an ulcer diverticulum or neoplasm of the gastrointestinal tract into the retroperitoneal space

One of the clinically more important and serious causes of mediastinal emphysema is perforation of the esophagus and trachea

Rupture of these organs has been observed in penetrating and non penetrating injuries of the chest More commonly it may be encountered in the course of endoscopic examinations Pneumomediastinum may be caused by the presence of foreign bodies or develop in the course of their removal

Of utmost clinical significance is the development of pneumomediastinum in spontaneous rupture of the esophagus In this clinical entity rupture of the esophagus occurs in the absence of known pre existing disease of this viscus In its clinical symptomatology this syndrome may show marked resemblance to other acute thoracic or abdominal conditions such as myocardial infarction acute pancreatitis or perforated peptic ulcer

Though the exact mechanism of esophageal rupture is not fully understood it is believed that sudden forceful distention of the lower esophagus in conjunction with disturbed motor function are important contributing factors With few exceptions this catastrophic event has occurred in the course of pronounced vomiting Alcoholism and cerebral disease are considered to be predisposing for the development of this entity

The tear in the esophagus is almost always found in its lower third anteriorly or on the left side It measures usually from a few millimeters to several centimeters in length Rapid outpouring of gastrointestinal contents into the mediastinum or pleural spaces leads to the development of pneumomediastinum pleural fluid formation and basilar pulmonary infiltration

Pneumomediastinum may develop subsequent to dissection of air from the interstitial spaces of the lung into the mediastinum On the basis of experimental and clinical studies it is now widely accepted that rupture of the pulmonary alveolar bases may allow escape of air into the tissue spaces sheathing the blood vessels of the lung and bronchi These air bubbles may dissect toward the hilum of the lung and pass into the mediastinal compartments As the tension of air in the mediastinum



Fig 41 Mediastinal emphysema following an attack of bronchial asthma in a young child Air has dissected throughout the entire mediastinum sharply demarcating the mediastinal structures

rises secondary rupture into pleural cavities may occur leading to the development of pneumothorax

Macklin has carried out detailed studies of the normal anatomic relationship of the pulmonary alveoli and their underlying stroma Several factors may alter the normal anatomic relationships to such an extent as to permit escape of air from the alveoli into connective tissue spaces of the lung Overdistention of the air spaces of the lung on one side and reduction of the vascular lumen on the other side may create abnormal pressure gradients between the alveoli and interstitial spaces of the pulmonary parenchyma These pressure differences may cause a break of the alveolar bases with the development of pulmonary interstitial emphysema

According to Macklin the most common conditions which predispose to pulmonary interstitial emphysema and pneumomediastinum are the following

1 Atelectasis of the lung associated with overdistention of remote or adjacent pulmonary parenchyma

2 General overinflation of the lungs with or without increased intra alveolar pressure

3 Decreased pulmonary blood flow especially in conjunction with hyperinflation of the lungs

Obviously there are large numbers of pulmonary disorders in which these predisposing factors are prevalent Foremost among these conditions are atelectasis pneumonia pulmonary abscess and generalized or localized pulmonary emphysema Other causes of acute mediastinal emphysema are sudden compression of the chest anesthesia under positive pressure and resuscitation Pneumomediastinum is known to have occurred following near physiologic maneuvers such as heavy straining and intense coughing

It is noteworthy that in infancy and early childhood mediastinal emphysema is not infrequently detected in the course of various respiratory disorders It has been postulated that the pulmonary alveolar bases



Fig 42 Spontaneous mediastinal emphysema A 29 year old physician noted sudden onset of moderately severe substernal pain while walking up stairs Discomfort persisted for 3 days The air in the mediastinum disappeared within a very short time

are particularly vulnerable in these age groups leading more often to the described train of events Thus pneumomediastinum and pneumothorax are commonly observed in infants with hyaline membrane disease and in the fetal aspiration syndrome

The clinical syndrome of spontaneous mediastinal emphysema deserves special consideration In this clinical entity usually in the absence of any known contributing factor pneumomediastinum develops with striking symptoms and signs Sudden substernal pain often requires differentiation from angina pectoris A crunching or crackling sound in the mediastinum is a most characteristic sign in this condition Most authors agree that interstitial pulmonary emphysema is a causative factor in the pathogenesis of this condition

On rare occasions gas formation in bac

terial infections may be sufficiently marked to lead to the accumulation of numerous small gas bubbles in the tissue spaces The infection may be primarily localized in the mediastinum or may have originated in the neck and spread into the thorax In this respect this type of mediastinal emphysema has considerable resemblance to other emphysematous infections occurring in various body parts and organs

Radiologically, a collection of air in the mediastinum may be recognized on frontal views by a unilateral or bilateral bulging of the mediastinal pleurae These septa are delineated as delicate lines which are separated from the mediastinal organs by a band of increased translucency

On the right the septum may extend from the soft tissues of the neck to the right heart border On the left the septum may be seen in the region of the large vessels and the aortic knob reaching towards the diaphragm along the left heart border In the lateral projection air in the mediastinum may accentuate the contour lines of the mediastinal structures Thus the anterior surface of the heart and aorta and the posterior aspect of the sternum may be demonstrated with unusual clarity

It is noteworthy that small amounts of air in the mediastinum may not be recognized or may be detected only with great difficulty Under these circumstances the frontal or lateral projections alone may show air in very limited segments of the mediastinum In many instances mediastinal emphysema will lead to emphysema of the soft tissues of the neck which may be radiologically more pronounced than the air collection in the thorax

In spontaneous rupture of the esophagus pleural effusion and pneumothorax are important associated radiologic features These complications are usually found on the left less commonly bilaterally and very rarely exclusively on the right side Escape of gastric contents into the mediastinum may cause mediastinal cellulitis and basilar pulmonary infiltration

In those instances in which mediastinal emphysema is caused by rupture of the trachea or esophagus the site of perforation



Fig 43 Severe mediastinal emphysema in a 5 year old boy with rubeola pneumonia The emphysema surrounds the mediastinal organs and extends into the soft tissues of the neck There was moderately severe dyspnea present

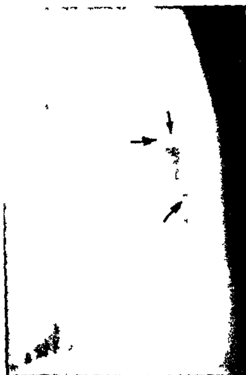


Fig 44 Spontaneous substernal mediastinal emphysema This 36 year old male patient developed a sudden pain in the suprasternal notch which gradually increased in severity A coarse grating sound was heard over the sternum The air was visible only in the lateral projection (arrows)



Fig 45 Spontaneous rupture of the esophagus A 57 year old male patient developed sudden onset of abdominal and left lower thoracic pain Air in the mediastinum and soft tissues of the neck (arrows) is an important diagnostic feature of this condition Note bilateral basilar pneumonitis and atelectasis and left pleural effusion

may be demonstrated by the administration of suitable contrast media

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Chapter 4

MEDIASTINITIS

MEDIASTINITIS has been defined by Kornblum and Osmond as inflammation of the loose mediastinal areolar tissue and its blood lymph and nerve supply The inflammation may be acute or chronic diffuse or localized suppurative or nonsuppurative Acute and chronic localized suppurations are termed abscesses diffuse suppurations phlegmons In the majority of instances mediastinitis can be controlled by antibiotics and external drainage and so is of less importance today than formerly

The causes for mediastinitis are numerous Cases may result from direct trauma (gunshot wounds stab wounds fractures surgical procedures) extension from inflammation of mediastinal structures (lymph nodes pericardium trachea esophagus) extension from perforation of mediastinal structures (esophagus trachea) and extension from inflammations in areas adjacent to the mediastinum (neck lungs pleura thoracic vertebrae retroperitoneal space)

Paravertebral abscesses which bulge into the mediastinum and which are almost always secondary to spinal osteomyelitis are discussed in the chapter dealing with lesions of bone and cartilage

The pathways through which inflammation spreads from the mediastinum to several adjacent areas (and vice versa) have occupied the attention of investigators for a long time Several studies dealing with this aspect of mediastinitis are mentioned in the references Most secondary types of mediastinitis spread downward from the neck and in doing so spread into the superior and posterior aspects of the mediastinum—not into the anterior compartment because of the structure of the fascial planes

Radiologically, mediastinitis presents a variable picture In mild cases of diffuse inflammation there may be no identifiable changes in the appearance of the mediastinum at all The borders will appear normal and the contours sharply defined With more advanced inflammations however there is likely to be diffuse widening poor definition of the borders (because of inflammation in the adjacent tissues) and even poor definition of the hilar shadows

With formation of an abscess by localization of a suppurative process the inflammation assumes more the appearance of a mediastinal mass Most abscesses are located either in the superior or posterior aspects of the mediastinum They cause unilateral or bilateral bulging of the borders They tend to be more commonly on the right than on the left They may displace the trachea or esophagus to one side or the other forward or backward Abscesses occur only rarely in the lower half of the mediastinum

Since lesions in the neck are important sources of mediastinal inflammation adequate studies should be made of this region Such examinations should include the following Frontal lateral and oblique film studies using either normal or soft tissue techniques of exposure fluoroscopy and contrast media studies of the esophagus Soft tissue emphysema either localized in pockets or extending diffusely along the muscle planes usually indicates perforation of one of the air containing structures (pharynx larynx trachea or esophagus) Increase in the distance between the posterior pharyngeal wall and the interior border of the cervical vertebrae identifies a retropharyngeal abscess Opaque foreign

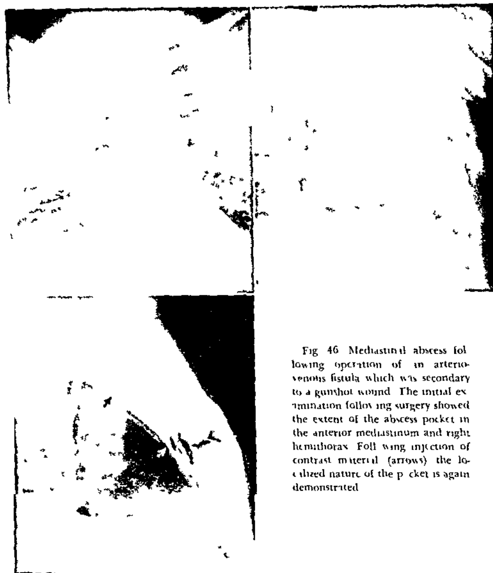


Fig 46 Mediastinal abscess following operation of an arteriovenous fistula which was secondary to a gunshot wound The initial examination following surgery showed the extent of the abscess pocket in the anterior mediastinum and right hemithorax Following injection of contrast material (arrows) the localized nature of the pocket is again demonstrated



Fig 47 Mediastinal abscess secondary to a cervical abscess This 50 year old patient swallowed a beef bone 5 days previously The bone lodged in the cervical esophagus perforated it and produced an extensive abscess The lateral view of the neck discloses the bone in the lower cervical area extensive soft tissue emphysema and forward displacement of the pharynx larynx and trachea The frontal chest film shows extension of the abscess into the superior mediastinum At esophagoscopy large quantities of yellow pus were obtained The bone was dislodged and passed into the stomach The mediastinum was drained Courtesy of George S Roach Jr M D and William W Bryan M D Atlanta Georgia



Fig 48 Mediastinal abscess secondary to a breakdown of tuberculous lymph nodes This 24 month old female had clinical findings of progressive tracheal obstruction which eventually required an emergency thoracotomy and tracheostomy Courtesy of Francis H Cole M D Memphis Tenn

bodies (bones and metallic objects) retained in the cervical esophagus can often be seen on appropriate studies

Examination of the entire esophagus is extremely important primarily because the underlying etiology of the mediastinitis is frequently an esophageal lesion Foreign bodies in the mediastinal segment of the esophagus are usually difficult to identify mainly because of the conflicting shadows of the lungs and bony structures of the thorax Fish and chicken bones can sometimes be identified by having the patient swallow a small pledget of cotton soaked in a contrast media and observing where the contrast media is arrested in the esophagus When perforations are apparent or suspected it is probably better to use a water soluble contrast media or iodized oil rather than a barium mixture

When the mediastinal mass is paravertebral in position the possibility that it is an abscess derived from a primary process in the thoracic spine should be borne in mind this is discussed more fully under the heading of paravertebral abscesses in the chapter dealing with lesions of bone and cartilage

Mediastinal emphysema similar to cervical emphysema is usually the result of perforations of the esophagus trachea or one of the bronchi This process has been described in detail in the chapter on emphysema and the reader is referred to that discussion In a rare instance mediastinal emphysema can be attributed to bacterial gas formation

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Fig 49 Localized abscess of the mediastinum secondary to esophageal perforation The elongated oval shaped abscess displaces the esophagus anteriorly



Fig 50 Mediastinal abscess secondary to esophageal perforation This examination discloses multiple air fluid levels and contrast media in the lower part of the abscess pocket There is extension into the right pleural space Courtesy of M Bedford Davis Jr MD and Steven G Cline MD Atlanta Georgia



Fig 51 Mediastinitis in a 47 year old male In a few days period the patient successively had a history of a molar tooth abscess Ludwig s angina induration and swelling in the anterior part of the neck and substernal pain Physical examination disclosed a friction rub with cardiac pulsations A chest examination revealed mediastinal emphysema Esophageal studies were normal Despite tracheostomy mediastinotomy and drainage of a submental abscess the patient died Post mortem examination disclosed massive necrosis of the mediastinum secondary to infection The mediastinal emphysema was evidently the result of bacterial gas formation since there was no perforation of a hollow viscus detected at autopsy

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Chapter 5

HEMOMEDIASTINUM

HEMOMEDIASTINUM has attracted little attention in the clinical literature. This condition should deserve more emphasis as its manifestations may be sufficiently severe to require surgical intervention such as decompression of the mediastinal structures. There is little doubt that hemorrhage into the mediastinum often remains unrecognized, as minor degrees of mediastinal enlargement may be difficult to recognize on radiologic and clinical examination.

Hemomediastinum may develop as a complication of various disease processes affecting the large vessels, such as rupture of a syphilitic or dissecting aneurysm of the aorta or its branches. A more unusual cause of mediastinal hemorrhage is the development of mediastinal hematoma in patients with uremia, typhoid fever and hemorrhagic diathesis. Capps has described the collection of a mediastinal hematoma in a patient with spontaneous hemorrhage originating from a parathyroid tumor. Under these circumstances the underlying primary pathology is usually in the foreground of the clinical picture though the mediastinal hemorrhage may have profound consequences.

The large majority of cases of hemomediastinum is post traumatic in origin. Fre



Fig. 52. Hemomediastinum in a patient with dissecting aneurysm. The frontal view prior to onset of symptoms shows a tortuous aorta without widening of the right paratracheal space. The outer contour of the right tracheal wall is well defined. Following the development of dissecting aneurysm a right paratracheal soft tissue mass appears obscuring the outer contour of the trachea. On autopsy this density was found to be a hemomediastinum. The dissecting aneurysm had ruptured into the pericardium.



Fig 53 Mediastinal hematoma following steering wheel injury of the sternum The superior mediastinum is widened as the result of mediastinal hemorrhage



Fig 54 Paraspinal hematoma This hematoma developed as the result of compression fractures of the sixth and seventh dorsal vertebrae



Fig 55 Hematoma of the mediastinum following stab wound injury of the bifurcation of the right innominate artery

quently mediastinal hematoma results fro external injury to the thorax particularl the anterior chest wall (steering wheel ac cident) One of the most common causes o mediastinal hemorrhage is a perforatin wound leading to the laceration of one of the large vessels The accumulation of blood and plasma in the mediastinum following surgical procedures in the region of the neck and mediastinum is commonly observed This type of fluid collection in the mediastinum is often noted following surgical procedures such as removal of mediastinal tumors correction of vascular malformations of the mediastinum or sympathectomy

Most often the hemorrhage is believed to stem from the venous system of the mediastinum Occasionally however the hemorrhage may be arterial in character particularly in the presence of penetrating chest wounds or rupture of an aortic aneurysm



Fig 56 Localized mediastinal hematoma following surgical correction of coarctation of the aorta (bedside roentgenogram) Several weeks later definite regression of the hematoma was noted



Fig 57 Hemomediastinum secondary to gunshot wound of the thorax The initial roentgenogram in frontal projection shows the bullet at the superior aspect of the hematoma Two weeks later the hematoma has almost completely disappeared Courtesy of S Krantz M.D. VA Hospital Atlanta Georgia



Fig 38 Large slightly lobulated mediastinal hematoma one day following surgical exploration of the mediastinum for parathyroid adenoma The hemomediastinum simulates a mediastinal tumor

The rapid accumulation of blood in the mediastinum may produce a tamponade like effect on the mediastinal structures resulting in venous distention cyanosis and dyspnea Blood loss may be sufficiently severe to lead to the development of shock

Radiologically, a small hemomediastinum may be difficult to detect on examination This applies particularly to those cases in which the radiologic examination is carried out in supine position at short focus film distances In early cases a slight obliteration of the individual contour lines of the mediastinal structures will be noted Larger degrees of hemomediastinum will cause a straightening of mediastinal borders in the sagittal projections With more pronounced hemorrhage a distinct convex smooth or even lobulated bulging of the mediastinal septa on one or both sides may be recognized Frequently the hematoma may dissect in o the lower portion of the mediastinum by gravitation

In the lateral view mediastinal hematoma may be detected as a bulging shadow along the posterior aspect of the sternum provided that this structure forms the base of the hemorrhage In trauma of the spine a bulging unilateral or bilateral paravertebral soft tissue mass is frequently recognized in the antero posterior projection In many instances the radiologic appearance of hematoma resembles that of other types of mediastinal mass formation For this reason careful correlation of radiologic findings and clinical findings appears necessary in this condition It is noteworthy that mediastinal hematoma may be absorbed rapidly

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Chapter 6

TERATOMAS

THE TERATOMA according to Willis is a true tumor or neoplasm composed of multiple tissues foreign to the part in which it arises It is not simply a malformation for unlike a malformation it possesses the powers of progressive growth

In the past the teratomas have been divided into epidermoids dermoids and teratomas depending on the number of germinal layers the lesions seem to possess But in recent years it has been conceded that practically all if not all of these tumors contain

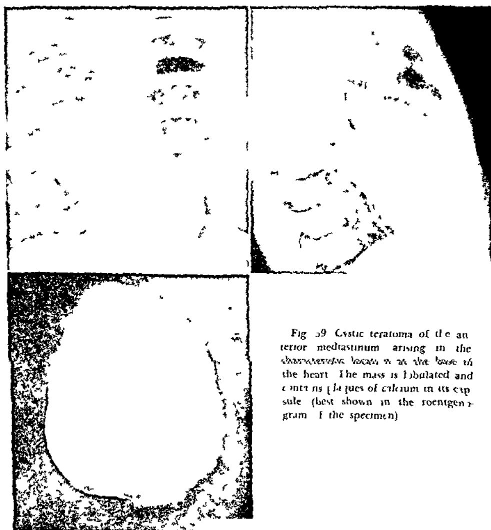


Fig 59 Cystic teratoma of the anterior mediastinum arising in the characteristic location at the base of the heart The mass is lobulated and contains plaques of calcium in its capsule (best shown in the roentgenogram of the specimen)



Fig 60 Teratoma dermoid type which ruptured spontaneously into the right pleural space A frontal roentgenogram of the chest discloses a mediastinal mass whose left border is visible against the lung but whose right border is obscured by pleural fluid Following the removal of 500 cc of fluid a second examination with heavier exposure discloses the true size of the mass and the presence of a tooth (arrow) within it From Hanten *et al* Spontaneous rupture of mediastinal dermoid cysts into the pleural cavity *Radiology* 64 548 1955

all three layers even though they may be quite variable in degree and that all three layers can be identified if thorough microscopic studies are made Qualifying adjectives for these teratomas have been suggested by Harrington to denote the tumors characteristics such as teratoma cystic type or teratoma dermoid type

The origin of the teratomas is not definitely known Willis discards the theory that they represent distorted fetuses for the following reasons They do not occur at the common sites of parasitic twins They often arise at a stage of development when their bearers have no mature germ cells capable of parthenogenesis They show no signs of a vertebral axis or of regional relationship of parts They are capable of independent progressive growth Rather the author believes that teratomas may represent areas of tissue which during early embryonic development escaped from the action of the primary organizer (i e the growth hormones which determine the orderly se



Fig 61 Chorionepithelioma of the mediastinum in a 27 year old male The diagnosis was originally made from a liver metastasis removed during surgery for an acute abdomen Assays ... amounts of ... evidence of tumor From Shlim N and van Brown D Extragenital chorionepithelioma in the male *Surgery* 28 755 1950



Fig 62 Huge teratoma in a child which apparently arose in the anterior mediastinum but which has grown so large that it extends from one lateral chest wall to the other and posteriorly to the level of the thoracic spine The tumor was successfully removed

Fig 63 Benign teratoma of the anterior mediastinum The mass is adjacent to the ascending aorta and an angiocardiogram was necessary to rule out an aneurysm



Fig 64 Probable teratoma of the anterior mediastinum This lesion which contained bone was unusual in that all sections of the tumor examined microscopically disclosed only the picture of a bronchogenic cyst The roentgenogram of the specimen shows the bony structures

quences of normal development and the mutual influences of growing plastic tissues on one another)

The *most common locations of these* tumors are the ovaries testes mediastinum and retroperitoneal space When in the mediastinum they are almost invariably in the anterior section just under the sternum and at the level where the great vessels join the heart at its base occasionally they are located intrapericardially Rarely these tumors are found in the posterior mediastinum Blades collected only 3 in 233 cases Occasionally they are dumbbell in shape with an extra sternal mass on the anterior chest wall connected with the mediastinal portion by a band of tissue penetrating the sternum

Grossly the mediastinal teratomas are smooth in outline The benign types predominate in cystic elements and for this reason are likely to be rounded or oval in shape Conversely the malignant forms are solid and are often lobulated Generally they are firmly attached to the surrounding structures notably the pericardium Both the malignant and benign forms may reach relatively large size—one of 6000 grams weight has been reported They usually increase in size with time the malignant forms being more rapid in growth In the *malignant forms squamous cell carcinoma* and adenocarcinoma predominate

Microscopically teratomas contain varying amounts of ectodermal mesodermal and entodermal structures including hair glands (mucous salivary sebaceous sweat) cartilage bone teeth intestine appendages and such tissues as muscle connective fibrous lipoid nerve thymic pancreatic and pulmonary

Spontaneous rupture into adjacent organs and spaces is not uncommon this may occur into the pericardium the heart aorta pulmonary artery superior vena cava an air passage or one of the pleural spaces Sudden death may result from this rupture

Symptoms result from compression and invasion of surrounding structures *from inflammatory* changes within the masses and from ruptures into adjacent structures Expectoration of hair is an interesting *pathognomonic sign*

Chorionepithelioma a highly malignant teratomatous tumor is usually encountered in the uterus of the female or the testis of

the male but occasionally is thought to be primary in the mediastinum. The difficulty in establishing an undeniable diagnosis of "primary" rests in the fact that proven mediastinal chorionepitheliomas may be secondary to tumors of the uterus or testis which have completely disappeared or at least have left only a residual scar which may be difficult to find on microscopic examination. When in the mediastinum these tumors are located anteriorly, may become quite large and invasive, and usually metastasize widely to the lungs, liver, brain and other organs. In both males and females they are frequently accompanied by bilateral gynecomastia and the presence of chorionic gonadotropin in the urine.

Radiologically, teratomas in practically all instances are seen as masses in the anterior mediastinum, particularly in the areas where the pulmonary artery and aorta arise from the heart. They may cause either unilateral or bilateral widening of the mediastinum. In their usual location they are prone to simulate cardiovascular abnormalities such as cardiac enlargement, ventricular aneurysm, and dilatation of the pulmonary artery, the aorta, or the adjacent large veins. In these instances fluoroscopy and film studies in multiple projections are important, and even with these a differential diagnosis between a mass formation and a vascular abnormality may be impossible. In such cases angiocardiographs may be necessary.

An occasional teratoma is located in the posterior mediastinum, but in this location it is not likely to be considered in a differential diagnosis unless it contains some of the characteristic components described below.

Teratomas of the mediastinum may contain teeth or other bony structures; such structures may not be apparent on routine chest roentgenograms, and heavily penetrated films or Bucky roentgenograms may be necessary for their identification. The presence of such parts indicates the diagnosis of teratoma. They may also contain plaques of calcium in the wall or compartments, but these alone do not help in their identification, since other anterior mediastinal tumors, such as thymomas, may contain similar calcium deposits.



Fig. 65. Teratoma, dermoid type, containing fatty material which layers out at the superior aspect of the lesion in these examinations made with the patient in upright and in right lateral decubitus positions. (From Phemister et al.: A roentgenologic criterion of dermoid cyst. Am. J. Roentgenol. 36:11, 1936.)

Phemister et al. have pointed out that the fatty material contained in the cystic forms of the teratomas may be visible as a

floating layer in the tumor in films made with the patient upright or in a decubitus position This finding is highly suggestive of the diagnosis of teratoma

Air fluid levels within teratomas indicate an acquired communication with one of the air passages Such a rupture may also produce a pneumonitis

The presence of fluid in one or both pleural spaces may indicate a rupture of a cystic type of teratoma The fluid can be so extensive that the tumor in the mediastinum is completely obscured

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Chapter 7

LIPOMAS AND LIPOSARCOMAS

LIPOMAS ARE benign tumors composed of adult fat cells these tumors tend to be lobulated well encapsulated and not very adherent to adjacent structures They may reach huge proportions (a 17 5 pound tumor of the mediastinum has been reported) before discovery Most of them arise in the anterior mediastinum and in their growth tend to broaden the mediastinum bilaterally Being soft they grow in the direction of least resistance Most are completely confined to the mediastinum but the occasional one may extend into the neck (cervicomediastinal) or may be dumbbell in shape with an extra mediastinal portion in the soft tissues of the thorax which is connected to the mediastinal component by an isthmus True lipomas in rare instances may arise within the involuted thymus gland of the adult which contains large amounts of fat

Hibernomas are unusual types of lipomas originating from homologues in man of hibernating glands Occasionally one of these occurs in the mediastinum although the usual site is in the interscapular region

Liposarcomas are malignant tumors of the lipoblast these usually consist of lipoblasts in different stages of development—from anaplastic to adult cells Unlike lipomas they are not proportionately greater in the anterior mediastinum rather they are more likely to be posteriorly placed And unlike lipomas they are usually discovered before they reach large size this is because of their tendency to produce symptoms They are lobulated and encapsulated but have a strong tendency toward invasion of adjacent structures They may suddenly accelerate in growth after slow growth for a long period Recurrence is frequent and metastases occur principally to the lungs liver and central nervous system this is considered to be a late manifestation

A patient with a liposarcoma in the mediastinum may have an associated liposarcoma elsewhere such as in the soft tissues of the thigh or gluteal region peritoneal cavity or retroperitoneal space Storey and Knutson have reported a rare combination of liposarcoma and lipoma occurring simultaneously in the mediastinum In a case reported by Heinemann and Lehman multiple sections through a mediastinal mass showed only liposarcoma but widespread metastases were those of a mixed mesodermal tumor (mesenchymoma) these tumors are discussed in more detail in another section of this book

Other mass lesions of the mediastinum which contain fat are epipericardial fat pads omental herniations through the foramen of Morgagni and dermoid types of teratomas All of these lesions are discussed elsewhere in this monograph

Radiologically lipomas are principally located in the anterior mediastinum and are usually quite large in size when initially discovered Their soft consistency allows growth in the course of least resistance and for this reason they may be somewhat bizarre in shape Those which grow around the heart may resemble pericardial effusions and on more than one occasion needle aspirations of these have produced fat Thorough roentgen study should make a differentiation between mass formation and fluid noting particularly any notches which might be present where the tumor and the heart meet this part of the examination is best carried out by fluoroscopy and spot films in multiple projections

The authors have personally observed one case of a lipoma which resembled an elevated diaphragm and only by examina

in either the lipomas or liposarcomas of the mediastinum

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Chapter 8

FIBROMAS AND FIBROSARCOMAS

TUMORS OF FIBROUS tissue are exceedingly rare in the mediastinum They arise from mesenchymal tissues and microscopically are composed of fibroblasts and an abundance of collagen (Schlumberger) in this respect they are similar to fibromas and fibrosarcomas seen in many other parts of the body Some authors refer to these as pure tumors in order to distinguish them from the more common neurofibromas which contain elements of nerve tissue

The reported cases of fibrous tissue tumors have been in various parts of the mediastinum seemingly having no predilection for one area or the other They have been reported as arising in the wall of the upper esophagus attached to the ribs spine sternum pericardium aorta and other adjacent soft tissues Their point of origin may be difficult to identify when they reach large size At least one reported case has shown multiple masses in the mediastinum—one anteriorly and one posteriorly

Grossly they are rounded or ovoid smooth walled and encapsulated The occasional fibroma may contain cystic components Occasionally they are lobulated Calcium deposits have not been reported in them They are often silent in their growth and reach large size before discovery one reported tumor weighed 1 972 grams Harrington reported a large 630 gram fibroma this tumor seemed to arise



Fig 69 Fibroma of the posterior mediastinum The mass is located behind the right hilum This lesion was diagnosed pathologically as a pure fibroma not one of neurogenic origin From Fouche J W Primary intrathoracic non pulmonary tumors *Am Surgeon 21* 909 1955

elements (liposarcoma chondrosarcoma and osteoid tissue) The mesenchymomas are discussed in more detail in another section of this book

Radiologically, these tumors of fibrous tissue origin appear as rounded ovoid or lobulated masses in the mediastinum The majority that have been reported have been relatively large in size In these cases it is often difficult to determine their site of origin

Practically all of those which have been reported in the literature have projected to one side only even though exceedingly large in size In these latter cases they usually cause marked compression of the *lung and displacement of the normal medi* astinal structures to the contralateral side

The mediastinal borders in general are sharply defined where the mass is located When the mass fills the entire hemithorax its entire circumference may rest against tissues of equal density and thus all borders are obscured

Their growth patterns may be quite striking radiologically and they should be particularly considered in a differential diagnosis when the following is apparent (1) The mediastinal mass has been known to be present for a period of time and has suddenly begun to enlarge (usually the result of sarcomatous degeneration in a pre existing fibroma) (2) The mediastinal mass is exceedingly large in size this latter point is particularly important if there have been few or no symptoms

Esophageal studies are of importance for several reasons The type of esophageal displacement if present may help in establishing the site of origin of the mass The characteristics of the esophageal deformity may aid in determining whether or not the mass arose in the wall of the esophagus In the case of esophageal fibromas the radiological findings are similar to the other primary benign lesions of the esophagus such as *the leiomyomas Primary fibrosar* comas of the esophagus according to Buckstein may be indistinguishable in appearance from the more common adenocarcinomas of that structure

The pleural effusions which accompany the benign and malignant types of fibrous tissue tumors of the mediastinum may tend to obscure the primary lesion particularly *if the examinations are made with the pa* tient in upright position in these cases further information can be obtained by fluoroscopy recumbent and decubitus films and possibly tomograms The mass itself *may resemble pleural fluid in instances* when it completely fills the hemithorax or when it almost does so and its superior border is relatively flat In these cases a radiological study with the patient in var*ious positions and a study of the com* pressed lung may aid in the differentiation between fluid and solid tumor

Angiocardiography may be necessary when fibromas and fibrosarcomas are located around the heart or the larger vessels in the mediastinum in order to differentiate these tumors from vascular lesions

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Chapter 9

MESENCHYMOMAS

MESENCHYMOMAS, according to Stout, are mixtures of tumor forms derived from the mesenchyme and amalgamated into a single mass. They are probably the result of faults in the embryologic development of the mesenchyme. They may be benign or malignant.

These tumors are exceedingly rare in the mediastinum but do occur in that location (Heinemann; Hobbs). In the two cases described in this section, both were initially considered to be liposarcomas, but were later found to be mesenchymomas. The more common locations of such tumors are the urogenital tract and the breast.

Stout reported a series of eight malignant mesenchymomas, none of which arose in the mediastinum. Each of these eight lesions had cells of two or more of the following types: fibrosarcoma, myxoma, liposarcoma, leiomyosarcoma, rhabdomyosarcoma, osteogenic sarcoma, chondrosarcoma, reticulum cell sarcoma, hemangioma, hemangiopericytoma, and hemangioendothelioma.

Two of his cases showed 5 types of tissues, one 4 types, two 3 types, and three 2 types. Every tumor differed from every other one in its cellular components, not one of them could be fitted into the standard varieties of sarcoma, and each one would have to receive a different compound name if they were to be designated by a term recording all of the component parts.

Mesenchymomas often grow to very large size and have a very strong tendency to recur after removal. Their microscopic picture is very varied as would be expected. Metastases occur, but are a late manifestation of the disease; they have a predilection for the central nervous system, lungs, and liver (Hobbs).

The radiologic findings in mediastinal mesenchymomas must of necessity be based on only 2 cases, since these are the only ones the authors have found in the literature. Both of these were malignant tumors and were located in the anterior mediastinum. Both were initially considered to be liposarcomas by biopsy, but their subsequent course and findings at post mortem examination showed them to be mesenchymomas.

Heinemann and Lehman described a mesenchymoma of the anterior mediastinum in a 63 year old white female. Radiographs of the chest revealed a large anterior mediastinal mass. At thoracotomy the mass could not be removed entirely because its upper portion was intimately attached to the great vessels. Microscopic examination of multiple sections of the removed portions showed only liposarcoma. Later, a mass in the right deltoid region was removed and showed well differentiated fibrosarcoma. The patient died 3 years and 5 months after her original diagnosis, and post mortem examination revealed well differentiated liposarcoma in a large mediastinal mass, liposarcoma and fibrosarcoma in a clavicular mass, fibrosarcoma and osteoid tissue in subpleural pulmonary nodules, well differentiated fibrosarcoma in arm and thigh lesions, and fibrosarcoma, liposarcoma, and chondrosarcoma in the heart muscle. These authors concluded that while the primary tumor was conclusively proven to be a certain type of lesion, the appearance of the metastases proved it to be otherwise. They further pointed out that an unusual feature of this case was the rather "pure" character of the metastases and that very little liposarcomatous tissue was found in the secondary lesions—some



Fig 73 Mesenchymoma of the anterior mediastinum in a 63 year old female The case is described in the text This radiographic examination was made sometime after the thoracotomy and shows the lesion in the anterior mediastinum and in the left clavicle Courtesy of W L Lehman M D Portland Oregon

thing to be expected since that type of tumor is not so apt to metastasize

Hobbs described the case of a 58 year old white female whose chief complaints were those of weight loss chest pain dyspnea Chest roentgenograms revealed a large mass occupying the lower part of the right chest At thoracotomy this was found to be an anterior mediastinal tumor extending laterally between the upper and



Fig 74 Mesenchymoma of the anterior mediastinum showing the original tumor The case is described in the text From Hobbs A W Malignant mesenchymal tumor simulating liposarcoma originating in the mediastinum *J M A Georgia 44 444 1955*

middle lobes of the right lung The mass was removed its weight was 2 950 grams The pathological diagnosis was liposarcoma Two subsequent thoracotomies were done for recurrences She died of the disease 6 years and 4 months after its initial discovery Tissues taken from the mediastinum at the time of death were examined by several pathologists the opinions mentioned were recurrent thymoma fibrosarcoma rhabdomyosarcoma and mixed mesodermal tumor (malignant mesenchymoma) with myosarcomatous and liposarcomatous elements There were no metastases to distant structures in this case

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Chapter 10

TUMORS OF BLOOD VASCULAR ORIGIN

"TUMORS OF BLOOD vascular origin" is a term applied by Seybold *et al* to describe a group of lesions of the mediastinum which arise from the vascular mesenchyme. These authors found the following pathologic terms used in the original reports in 17 cases of this type which they collected: endothelioma, pleural endothelioma, malignant endothelioma, hemangiosarcoma, angioma, hemangioma, malignant hemangioendothelioma, and angiosarcoma.

All of these blood vascular tumors have the histologic picture of blood vascular spaces lined with endotheliomatous tumor cells. Some are quite simply constructed (such as the cavernous hemangiomas); others have a predominance of these cells but include other tissues of mesenchymal origin as well. At times phleboliths are contained within the tumors and, when present, are pathognomonic.

Histologically, these tumors may be benign or malignant. Some of the malignant types metastasize widely. Some of these tumors are well encapsulated and easily removed at surgery; others are so infiltrative and invasive that they cannot be eradicated entirely. For the most part they are discovered in adult life between the ages of 20 and 40.

These lesions may have their origin in the mediastinum and remain confined to that area, or may arise adjacent to the mediastinum and secondarily encroach upon it. Carlson and Adams reported a case which arose in the musculature of the thorax and extended from there into the posterior mediastinum and the extradural space; their patient had an associated hypertrophy of the upper ribs bilaterally. Adams and Bloch have reported coexistent tumors of this type in the mediastinum and skin. At times two or more tumors of blood vascular origin may occur in the mediastinum simultaneously; they are considered to be multiple primary growths—not single growths with metastatic spread.

These tumors are quite variable in size and may assume huge proportions. Even these may remain noninvasive. Blood vascular tumors may grow rapidly in children. Bergstrom reported relatively large tumors of this type in 2 newborns; one of these patients had progressive respiratory difficulty because of the mass, resulting in death on the twenty-sixth day of life; the other had a lesion which arose in the thymus and which ruptured into the right pleural space, causing death by hemorrhage on the thirty-sixth day of life.

Radiologically, tumors of blood vascular origin are located predominantly in the anterior mediastinum or in the anterior part of the superior mediastinum. In a series of 19 cases reported by Ellis *et al*, 17 were anteriorly and only 2 posteriorly. They may be located at any level, and in this respect are different from some other types of anterior mediastinal masses, such as teratomas and thymomas, which are usually (but not always) found around the base of the heart.

The borders of these masses against the air-filled lungs are usually sharply defined; those lying against the mediastinal structures of equal density are not discernible. Radiologically, it is usually impossible to say whether these masses are invading adjacent structures or not.

Phleboliths are contained in some of these lesions in the mediastinum and are similar to those contained in like tumors

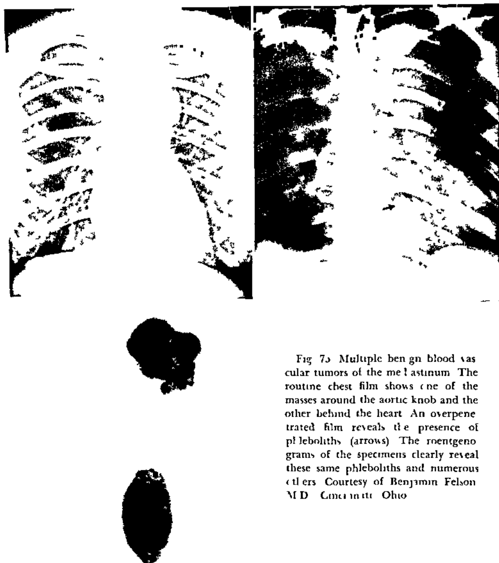


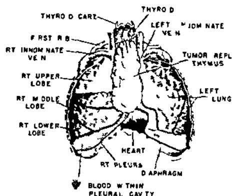
Fig 73 Multiple benign blood vascular tumors of the mediastinum The routine chest film shows one of the masses around the aortic knob and the other behind the heart An overpenetrated film reveals the presence of phleboliths (arrows) The roentgenograms of the specimens clearly reveal these same phleboliths and numerous others Courtesy of Benjamin Felson M D Cincinnati Ohio



Fig 76 Benign blood vascular tumor in a 40 year old female The frontal view discloses the right border of the mediastinal mass paralleling the right heart border the lateral view shows its posterior border distinctly This lesion involved the pericardium the right pulmonary vein and the inferior vena cava Excessive bleeding was encountered at operation The mass could not be removed Courtesy of Herbert M Olnick M D Macon Georgia



Fig 77 Benign blood vascular tumor in a 29 year old male who sustained 2 penetrating chest wounds during the war Radiologic examination several years later disclosed a left anterior mediastinal mass as shown here Venography revealed retrograde filling of a portion of the lesion from the left subclavian vein Surgery confirmed these findings and the mass was removed Histologic examination disclosed numerous small and large vascular spaces lined with endothelium Courtesy of Simon Krantz M D V A Hospital Atlanta Georgia



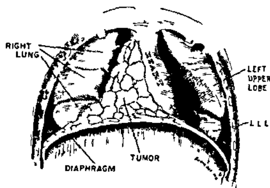


Fig 79 Benign blood vascular tumors *Sketches showing the post mortem findings in the mediastinum of two infants who died as a result of these tumors* From Bergstrom V W Hemangioma of mediastinum causing death in newborn *New York State J Med* 45 1867 Sept 1 1945

in the soft tissues of the extremities and other areas of the body these are rounded or oval shaped thrombi containing cal cium and are located in the vascular spaces When present they are highly char acteristic They may not be visible unless adequate studies are made with overpene trated films or with tomograms

Angiography has been used very little in the study of these lesions in the medi astinum As a general rule they do not opacify with the contrast media because of the lack of large vessels entering them

Fluid in the pleural space in association with a vascular tumor in the mediastinum may be the result of metastatic involvement (in the case of a malignant lesion) or rup ture of the primary lesion in the mediasti num (as happened in one of Bergstrom s newborns)

The occasional tumors of blood vascular origin which extend into the spinal canal from the mediastinum may produce neuro logical symptoms and in such cases my elography is helpful in determining the extent of the lesion Bony erosion has not been reported but bony hypertrophy (up per ribs bilaterally) was present in one such case reported by Carlson and Adams

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Chapter 11

LESIONS OF LYMPH VESSELS

FOR THE MOST part these are tumors and cysts of lymph vascular origin which are found at all levels in the mediastinum and usually in the anterior portion One rare form of cyst however is that arising from the thoracic duct in the posterior mediastinum because of the differences between this type of lesion and the others it is discussed separately in this text Lymph node lesions are described in a separate chapter

TUMORS AND CYSTS OF LYMPH VASCULAR ORIGIN

Tumors and cysts of lymph vascular origin are derived from the mesenchyme just as are those of blood vascular origin They are known by many names The benign forms are commonly referred to as lymphangioma hygroma cystic hygroma lymphatic cyst lymphogenous cyst cystic lymphangioma cavernous lymphangioma and chylous cyst The malignant varieties are described as lymphangio endothelioma and lymphangiosarcoma

These lesions may arise in the mediastinum and be wholly confined to it (mediastinal type) or may arise in the neck and extend secondarily into the superior mediastinum (cervicomediastinal type) The former is rare and is usually first discovered in adult life the latter is more common and is practically always first discovered in infancy because of the presence of a mass in the neck The origin of these masses is not definitely known Schlumberger states that the embryonic lymphatic sacs in the neck are the probable site of origin of the cervical and cervicomediastinal hygromas of childhood but there are no similar lymphatic sacs in the mediastinum during embryogenesis to explain the mediastinal lesions which are usually discovered at a later period He believes that some of the so called lymphangiomas may in fact be multiloculated thymic cysts in as much as these 2 lesions are microscopically indistinguishable at times

Grossly these lymphatic lesions vary from simple thin walled unilocular cysts loosely attached to surrounding tissues to multilocular tumors which are intimately associated with the surrounding structures such as the pericardium and great vessels These masses enlarge by a process of endothelial sprouting (Goetsch) the endothelial buds in their growth along tissue planes envelop such structures as vessels and nerves and marked adherence results between the



Fig 80 Lymph vascular tumor (the so called hygroma) in an infant cervicomediastinal in position The mediastinal portion has a smooth right border a lobulated left border The patient had a palpible soft tissue mass in the neck anteriorly which displaced the larynx and trachea to the right

Fig 81 Lymph vascular tumor of the anterior mediastinum invading the pericardium The lesion causes the heart to appear enlarged but an oblique spot film discloses a characteristic notch between the two structures aiding in the differentiation The lesion could not be completely removed



Fig 82 Malignant lymph vascular tumor of the anterior mediastinum This circumscribed mass was removed without difficulty

eventual tumor and the encompassed structures

The lesions are quite variable in size measuring from a few centimeters in diameter to masses filling a large portion of the chest. Their borders may be smooth or lobulated. In the cervicomediastinal types part of the lesion is within the neck and the remainder extends down into the mediastinum.

Microscopically these tumors and cysts are composed of cystic spaces lying within endothelium; their walls are variable in thickness. There may be considerable connective tissue stroma present. The fluid within the cystic spaces of these lesions is usually clear or amber colored, thin, low in protein content, and contains a few cells (chiefly mononuclears and lymphocytes). Occasionally the fluid may be turbid or hemorrhagic. At times instead of fluid the cyst may contain a thick gelatinous material.

The lymph vascular lesions confined entirely to the mediastinum may be located at any level, usually anteriorly; a posterior location is rare but does occur. The cervicomediastinal lesions cause swelling in the neck as well as mediastinal tumefaction.

Radiologically the mediastinal lesions of lymph vascular origin are seen as rounded, lobulated or irregular masses. The borders against the lung are sharply defined. The projection is usually to the right but occasionally it occurs to the left. The cervicomediastinal lesions extend for a variable distance into the superior mediastinum and will present similar borders either to one or both sides; the cervical part of the mass may be apparent on chest examination either as a soft tissue mass or by displacement of structures such as the larynx, pharynx, trachea or esophagus.

Both types are of homogeneous density. They evidently do not calcify. There may be compression and displacement of surrounding structures by both types depending on size, location and structure. Film

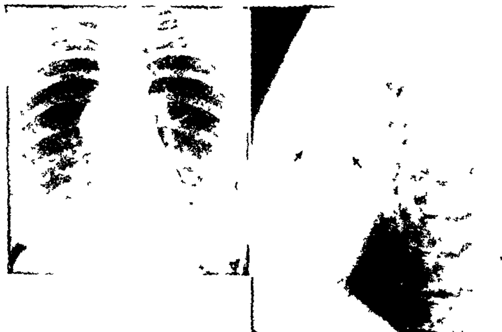


Fig. 88. Lymph vascular tumor of anterior mediastinum. The tumor has a lobulated margin best demonstrated in the lateral view (arrows). Courtesy of William Molnar, M.D., Columbus, Ohio.

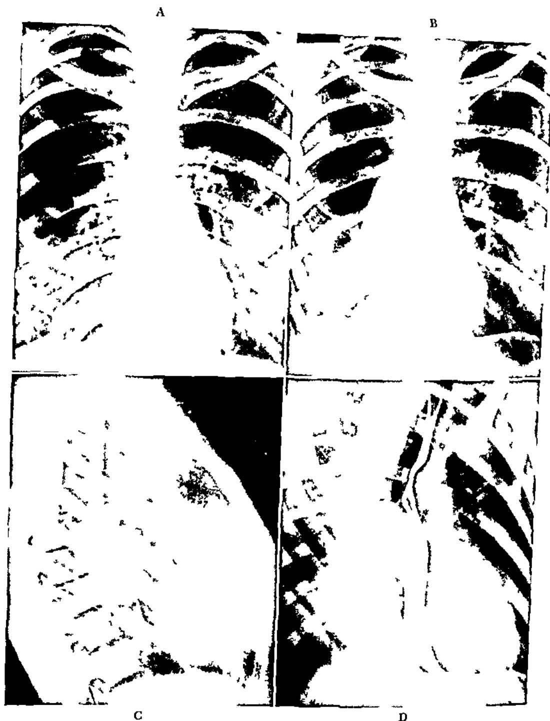


Fig 81 Thoracic duct cyst in a 20 year old college student Two years prior to hospital admission a chest examination (A) showed a slight widening of the mediastinum to the right at the hilar level Examination at the time of admission (B C and D) revealed an increase in the size of the mass and some associated changes in the right lower lobe At surgery the mass proved to be a cystic dilatation of the thoracic duct From inside the mass a cannula was slipped into the duct leading away from the cyst superiorly and an injection of contrast media was made a film (E) taken immediately afterward showed opacification of the thoracic duct From Emerson G L Supradiaphragmatic thoracic-duct cyst *New England J Med* 242 575 Apr 13 1950 Courtesy of Alfred J Kummer MD and Robert M Lowman MD New Haven Connecticut

studies and fluoroscopic observations in various projections may help in determining these relationships The mediastinal portion of a cervicomediastinal lesion may appear to diminish in size with expiration due to displacement of some of the mass into the neck (Gross *et al*)

Pleural fluid in association with a mediastinal lesion of lymph vascular origin may be on the basis of an accompanying chronic chylothorax

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Fig 84E

THORACIC DUCT CYSTS

Cysts of the thoracic duct are exceedingly rare only a few have been reported and most of these were discovered incidentally at post mortem examination

The thoracic duct is the main collecting trunk of the lymphatic system of the body It has its origin at the cisterna chyli which is located just anterior to the second lumbar vertebra the cistern is the main drainage channel for the lower part of the body The duct leaves the abdomen through the aortic hiatus of the diaphragm ascending in the midline of the posterior mediastinum between the aorta and the azygos vein Opposite the fifth dorsal vertebra it veers to the left passing behind the aortic arch and the mediastinal portion of the left subclavian artery then it passes into the left side of the neck where it empties into the venous system at the angle between the left subclavian and the internal jugular veins Generally it is tortuous and irregular in diameter At times it divides in the middle of its course into two channels which soon unite At other times it consists of multiple channels giving to the system a plexiform appearance The course of the thoracic duct is illustrated in the chapter of mediastinal anatomy

The few cases of thoracic duct cysts that have been reported in the literature have been reviewed by Emerson Some of these were unilocular others multilocular Some apparently arose secondary to lymphangitis All levels of the mediastinum were involved In Emerson's own case and in a similar one reported by Bakst the cyst was located in the posterior mediastinum just above the diaphragm and was unilocular During surgical removal the cyst when collapsed would refill with chyle from the thoracic duct which entered it inferiorly

In Emerson's case the thoracic duct continued off the superior side of the cyst in Bakst's none could be identified the cyst evidently being a blind pouch In each case removal of the cyst and ligation of the duct did not produce any harmful effects evidently there were collateral channels in the mediastinum

Radiologically, thoracic duct cysts are seen as posterior mediastinal masses broadening the borders to one or both sides Post mortem description in the older reported cases and radiological descriptions in the more recent ones would indicate that they may be quite variable in size single or multiple smooth or lobulated and be located at any level Evidently they do not contain significant calcium deposits and do not erode bone They may displace adjacent structures notably the esophagus When still growing interval films may show a progressive increase in their size

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Chapter 12

LESIONS OF THE LYMPH NODES

LYMPHADENOPATHY is one of the most common causes of mediastinal mass formation. Enlargement of mediastinal lymph nodes may be benign in character or may develop in the course of various types of malignant processes such as lymphoma, leukemia and carcinoma.

NON MALIGNANT LYMPHADENOPATHY

There are many benign diseases which cause enlargement of the mediastinal lymph nodes. Most often the mediastinal adenopathy is part of a disease process confined to the thorax. In other instances, however, mediastinal adenopathy is the result of a widespread involvement of the lymph nodes throughout the entire body.

Foremost among the non malignant types of mediastinal lymphadenopathy are those conditions in which the enlargement of the lymph nodes develops in the course of pulmonary or pleural infections. Mediastinal lymphadenopathy has been described in a large variety of bacterial, viral, as well as fungus diseases. A detailed description of these conditions would go beyond the scope of this monograph. The lymphadenitis may be acute or chronic and terminate in necrosis, fibrosis and calcification. In some instances the mediastinal lymphadenopathy is in the foreground of the clinical and radiologic picture inasmuch as the pulmonary focus of the infection may be minimal or difficult to recognize on radiologic examination. Occasionally extensive mediastinal lymph node enlargement leads to compression or erosion of adjacent structures such as the trachea, bronchi and large vessels. In mediastinal lymph node tuberculosis atelectasis, bronchiectasis and hemoptysis have been encountered as a complication.

There are other conditions in which mediastinal lymphadenopathy often arises in con



Fig. 8. Primary tuberculosis of the left lung and regional lymphadenopathy in a 9 year old boy. Note marked enlargement of the left prevascular chain (arrows). Following treatment there was marked regression of pulmonary disease and mediastinal lymphadenopathy.

junction with pulmonary disease forms an essential part of the radiologic diagnosis These include sarcoidosis erythema nodosum and various types of pneumoconioses The lymph nodes of silicosis often show peripheral calcification which has been described as egg shell in type Though this type of calcification is not pathognomonic for this entity it may be considered as characteristic It should be kept in mind that silicosis is frequently associated with infections such as tuberculosis and histoplasmosis which may contribute to the enlargement and calcification of the involved lymph nodes

In general the distribution of lymph node enlargement in the mediastinum in a large variety of infectious diseases does not assume a characteristic pattern It should be stated however that in sarcoidosis involvement of the mediastinal lymph nodes in the absence of intrapulmonary lymphadenopathy is very rare On the other hand involvement of the prevascular lymph node system in patients with lymphoma is commonly found in the absence of intrapulmonary lymph node enlargement The anatomic position of the lymph nodes and their involvement in a variety of diseases has been described in the chapter devoted to the anatomy of the mediastinal lymph nodes



Fig 86 Tularemia Pulmonary infiltration and enlargement of the right hilar and right paratracheal nodes in a 37 year old patient who had skinned wild rabbits prior to the onset of illness The agglutination tests were strongly positive for tularemia

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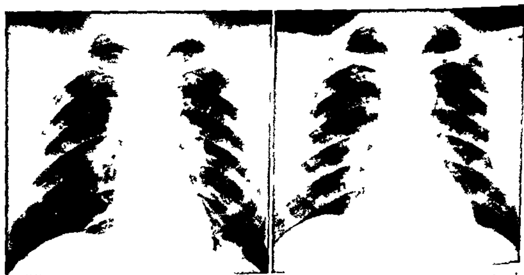


Fig 87 Sarcoidosis Marked enlargement of the intrapulmonary and mediastinal lymph nodes in a patient with sarcoidosis Reexamination after two years reveals marked regression of the lymphadenopathy

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LYMPHOMA

By definition lymphoma means tumor of lymphoid tissue By common usage lymphoma denotes a primary malignant disease of the lymphoid tissues of the body The term malignant lymphoma is also used Although lymphadenopathy is the most common manifestation of the disease lesions can occur anywhere in the body where there is lymphoid tissue

Lymphoma has been classified in several different ways One classification that has found wide acceptance is that of Gall and Mallory which is based on cytologic appearance these authors state that it is easily possible to divide the great majority of specimens into seven categories of malignant lymphoma as follows

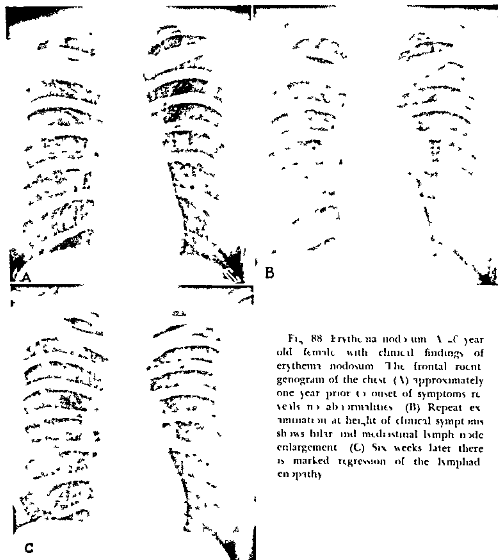


Fig 88 Erythema nodosum A 26 year old female with clinical findings of erythema nodosum The frontal roentgenogram of the chest (A) approximately one year prior to onset of symptoms reveals no abnormalities (B) Repeat examination at height of clinical symptoms shows hilar and mediastinal lymph node enlargement (C) Six weeks later there is marked regression of the lymphadenopathy



Fig 89 Silicosis Egg shell calcifications of mediastinal lymph nodes (arrows) in a patient with extensive pulmonary silicosis

(1) Stem cell lymphoma } Reticulum cell sarcoma
(2) Clasmatocytic lymphoma }
(3) Lymphoblastic lymphoma
(4) Lymphocytic lymphoma
(5) Hodgkin s lymphoma } Hodgkin s disease
(6) Hodgkin s sarcoma }
(7) Follicular lymphoma

The first four of these predominate in a single cell type The Hodgkin s lymphoma in contrast is predominantly polycellular having present neutrophiles eosinophiles lymphocytes plasma cells and fibroblasts as well as reticulum cells and some Sternberg Reed cells The Hodgkin s sarcoma is composed mainly of Sternberg Reed cells

Gall and Mallory believe that the terms lymphosarcoma and leukosarcoma should be discarded since these appear to represent transient phases of malignant lymphoma and do not constitute disease entities They further believe that the development of leukemia at some phase during a lymphoma s progress (a common occurrence) should be regarded as an incidental occurrence in the disease—simply an overt manifestation of the underlying process

As stated earlier lymphoma can invade any part of the body where there is lymphoid tissue The most commonly affected parts include the lymph nodes spleen liver lungs gastrointestinal tract genito urinary tract skin and bones The more disseminated the disease the shorter the period of survival

The mediastinum an area that is involved in about one half of cases contains several major groups of lymph nodes (see under Anatomy) Some or all of these may be involved in any given case the location and extent of involvement determining the appearance of the mediastinal mass or masses The mediastinal nodes are not a separate system of nodes but are merely one area richly interconnected with adjacent nodes in the lungs supraclavicular areas neck breasts abdomen and other areas

In some cases involvement of the mediastinum may be the presenting manifestation without evidence of other adenopathy Kasabach and McAlpin in a study of 7? cases of Hodgkin s disease having mediastinal involvement had 19 without adenopathy elsewhere at the time of the initial examination in these cases peripheral node enlargement became apparent on an average of 18 months later In their other 58 cases other areas as well as the mediastinum were involved at first examination

Jackson and Parker in their monograph on Hodgkin s disease make the following observations In 171 cases of Hodgkin s granuloma 90 had mediastinal or hilar adenopathy during the course of the disease Of these 90 cases 36 per cent had these lesions from the outset and another 38 per cent had them within one year In their cases of Hodgkin s sarcoma one third had involvement of the mediastinal lymph nodes at post mortem examination Pulmonary involvement was quite frequent in

both types These authors state that such facts indicate clearly the necessity for roentgenray study of the chest when the patient is first seen even though there are no symptoms or signs even remotely suggesting such lesions

Radiologically, lymphoma of the mediastinum produces no characteristic pattern that is diagnostic of the disease during the entire course of a single case there is likely to be considerable variation in the appearance of the lesion in the mediastinum The size distribution and extent of involvement of the lymphoid tissue are the determining factors

When the disease is minimal the mediastinal widening may be barely perceptible and limited to a single site This is especially true when the paratracheal nodes are enlarged and the lesion resembles the lateral border of the ascending aorta on the right or the aortic knob on the left or when the subcarinal nodes are enlarged and obscured by the heart in the frontal and by the hilar vessels in the lateral view

As the disease progresses the mediasti

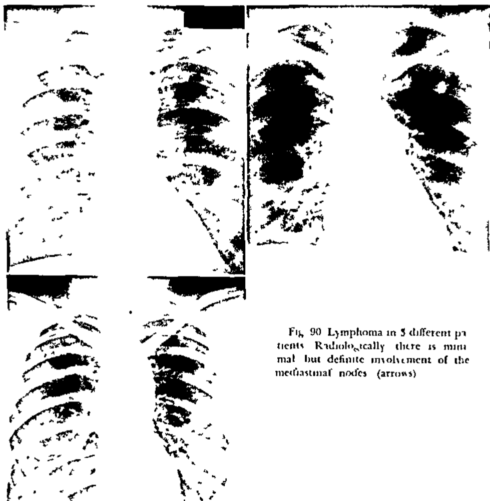


Fig 90 Lymphoma in 5 different patients Radiologically there is minimal but definite involvement of the mediastinal nodes (arrows)

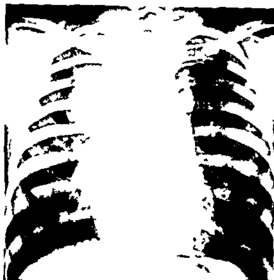


Fig 91 Lymphoma involving multiple groups of lymph nodes In such cases there is characteristically bilateral lobulated mediastinal tumefaction as shown here



Fig 92 Lymphoma predominantly involving the internal mammary chain of lymph nodes In this lateral examination of the chest the presternal edema and the retrosternal board like infiltrates are apparent

num enlarges further this may be unilateral in nature but is more likely to be bilateral Usually the borders are lobulated The involvement may be so great that the mediastinal pleura approaches or rests against the lateral thoracic wall pleura In such cases there is marked compression of pulmonary tissues

At times the chain of lymph nodes in the anterior mediastinum is predomi





Fig 93 Lymphoma with enlargement of the lymph nodes in the anterior mediastinum



Fig 96 Lymphoma involving the spine The routine chest examination in frontal projection discloses right pleural space fluid and a fusiform mediastinal mass suggesting lymphadenopathy A similar examination with heavier exposure discloses that the mass is actually a paravertebral extension of a lymphoma which involves at least 2 vertebrae and the adjacent ribs

ciated with mediastinal tumefaction it lends strong support to the diagnosis of lymphoma or leukemia

Mediastinal lymphoma is commonly accompanied by manifestations of the disease elsewhere in the chest In the lungs there may be hilar adenopathy (unilateral or bilateral) single or multiple peripheral masses of variable size diffuse linear or nodular densities and segmental consolidations At times the masses may cavitate At times fungus infiltrations accompany the pulmonary lymphoma Vieta and Craver state that isolated nodules of lymphoma in the lungs when they occur are almost invariably accompanied by other types of intrathoracic manifestations of the disease Pleural involvement with lymphoma often produces effusion Pleural masses if they reach sufficient size and are not obscured by fluid may be visible on the chest roentgenograms The bones of the thorax including the ribs sternum clavicles and dorsal spine may show lesions (Coles and Schulz) these may be osteolytic or osteoblastic single or multiple

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LEUKEMIA

Leukemia is a malignant disease of the hemopoietic organs affecting principally the bone marrow and the lymphatic and reticuloendothelial systems According to Ackerman and Regato the several types of leukemia may be classified as follows based on the cell of origin

(1) Myelocytic leukemia from the myeloblast

(2) Lymphocytic leukemia from the lymphoblast

(3) Monocytic leukemia from the monoblast

(4) Plasmacytic leukemia possibly from cells of the reticuloendothelial system

Each of these types may be further divided into acute and chronic types

Pathologic changes in leukemia may be of two types (1) Changes which affect the blood forming organs (particularly the bone marrow spleen and lymph nodes) and (2) changes due to infiltration These pathologic changes frequently involve many different organs and tissues of the body in one or both ways In this monograph however attention is directed primarily to leukemic involvement of the mediastinum (i e lymph nodes) and secondarily to other areas of the body which are radiologically demonstrable

Mediastinal adenopathy is usually a part of a generalized adenopathy involving many lymph node groups it is rare that the mediastinum is involved alone or that the disease is first discovered on a chest examination

The several types of leukemia vary considerably in their degree of lymph node involvement Acute and chronic lymphocytic leukemia almost constantly cause lymphadenopathy this adenopathy is usually generalized though the extent of enlargement in different groups of nodes is variable The mediastinal and retroperitoneal nodes are usually not enlarged at first but may show enlargement later The adenopathy in the lymphocytic type is a result of infiltration by small lymphocytes

In myelocytic leukemia adenopathy may be present particularly in the late



Fig 97 Chronic lymphocytic leukemia with adenopathy in the neck mediastinum and hilar areas all visible on a single chest examination

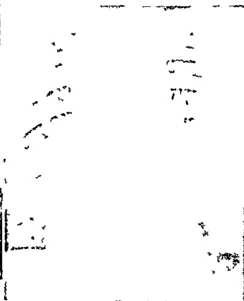


Fig 98 Monoblastic leukemia with enlargement of mediastinal and hilar lymph nodes



Fig 99 Acute lymphatic leukemia At the time the diagnosis was established a chest examination disclosed only questionable *mediastinal enlargement* *Three months* later the left sided lymph node groups were extensively involved

stages of the disease but it is usually mild and is a result of proliferation or infiltration *with the myeloid cells (Forkner)* Monocytic leukemia frequently has generalized adenopathy of moderate extent resulting from a replacement of the lymphoid cells by cells of the monocyte series

The involved lymph nodes even when greatly enlarged rarely show any tendency to invade surrounding structures The microscopic structure of the node is greatly altered particularly in the lymphocytic types where the normal architecture may be completely obliterated

Radiologically when there is leukemic involvement of the mediastinal nodes the chest examination will frequently show generalized broadening of the entire mediastinum both to the left and right sides In such cases the borders are usually straight or lobulated and sharply defined Isolated enlargement of nodes is infrequent being more common in malignant lymphoma than in leukemia Kirklin and Hefke however have pointed out that the radiologic appearance of generalized lymphadenopathy in the mediastinum is similar *in all the leukemias and lymphomas* and no differential diagnosis can be made between them

Cervical adenopathy is quite frequent in leukemia and lymphoma This may be visible on the chest examination *and aid* in differentiating the mediastinal adenopathy from other lesions which do not have accompanying cervical pathology

Pulmonary involvement in leukemia manifest itself in several ways *including* hilar adenopathy which may be unilateral or bilateral and pulmonary infiltrations which may be linear nodular patchy or miliary Pulmonary lesions are more common in lymphocytic than in myelocytic leukemia (Vieta and Craver) Pleural involvement frequently results in effusions when considerable fluid is present in the pleural space the entire mediastinal border may be obliterated

Hepatomegaly and splenomegaly are almost invariably present—a finding which

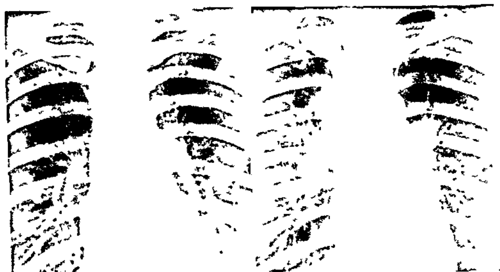


Fig. 100. Metastatic melanoma in a 40 year old female. Two examinations over a period of six and one-half months show development of metastases in the mediastinum and hilar regions.



Fig. 101. Mediastinal metastases from a small bronchogenic carcinoma of the left lung. The large mediastinal mass had resemblance to an aortic aneurysm. Courtesy of S. Krantz, M.D., V.A. Hospital, Atlanta, Georgia.

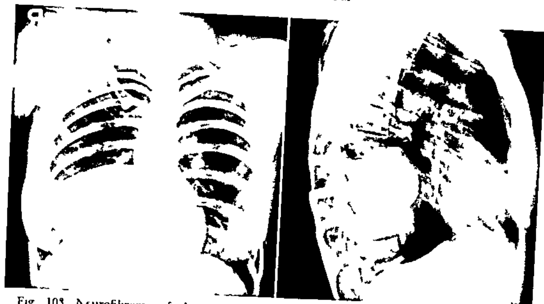


Fig 103 Neurofibroma of the posterior mediastinum showing slight lobulation

ones are solid but the larger ones may become partly cystic due to degeneration In the mediastinum they may grow to very large size They are benign and according to Stout it is questionable whether or not a malignant tumor ever develops from them

The neurofibromas contain not only the sheath cells but nerve elements as well They arise as solitary masses at times but more commonly are seen in the form of multiple neurofibromatosis (von Recklinghausen's disease) The solitary forms in the mediastinum resemble the neurilemmomas in their gross appearance

The malignant counterpart of the neurilemmoma is called the Schwannoma it arises from the Schwann cells of the nerve sheath These represent primary malignant tumors and not malignant degeneration in benign neurilemmomas They are sometimes called neurogenic sarcomas but this is not a preferred term (Stout) In the mediastinum they may be either encapsulated or infiltrative They have a tendency to metastasize freely through the blood stream

Most of the peripheral nerve tumors are located in the posterior parts of the mediastinum but the occasional one is further forward Tebow and Brown found 9 such tumors out of 217 mediastinal neurogenic tumors of all types the remaining 208 were located posteriorly

Grossly most of these peripheral nerve tumors are rounded or lobulated and well encapsulated Occasionally they may be infiltrative and involve surrounding struc

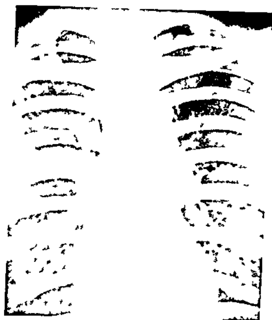


Fig 104 Schwannoma in a 30 year old male The poorly defined and irregular tumor is seen around the aortic knob and upper left hilum The left diaphragm is paralyzed as a result of the lesion At surgery the tumor was found to be invasive and could not be removed

Fig. 105. Neurofibroma in an atypical location. The chest examination discloses a mass located in the anterior part of the superior mediastinum. Courtesy of James G. Gabbard, M.D., Corpus Christi, Texas, and the Mallinckrodt Institute of Radiology, St. Louis, Mo.



Fig. 106. Neurofibroma of both vagus nerves confirmed by surgery. Courtesy of Harold O. Peterson, M.D., Minneapolis, Minnesota.



Fig 107 Large ganglioneuroma occupying the major portion of the superior mediastinum The tumor is deforming the left upper rib cage

tures They are usually unilateral and may reach very large size

Peripheral nerve tumors arising in the spinal canal may grow through the inter vertebral foramen into the posterior medi astinum The reverse may also be true Those arising in the intervertebral foramen may grow in both directions In all these instances they become hour glass or dumb bell tumors usually with the 2 parts being asymmetrical in size Not un commonly these tumors in the posterior



Fig 108 Incompletely differentiated ganglioneuroma of the superior mediastinum and lower neck (cervicomediastinal) When first seen this 8 month old patient had respiratory stridor a left Horner s syndrome and venous congestion of the left arm and shoulder A *needle aspiration of the tumor was done and was reported as* malignant tum r sugges tive of neuroblastoma The lesion was irradiated At 22 months of age the mass was sur gically removed and the microscopic examination revealed both mature and undifferentiated ganglionic cells

mediastinum cause bony erosion In the spine this may be in the nature of an enlargement of an intervertebral foramen erosion of one or more pedicles or erosion of one or more vertebral bodies In the ribs it may be simply deformity or erosion or combinations of the two

Pleural fluid occasionally accompanies these tumors occurring with the benign forms as well as the malignant Schwannomas Harrington reported one case of bloody pleural fluid associated with a benign neurofibroma

Tumors of the Sympathetic Ganglia These include the benign and malignant ganglioneuromas and the malignant sympathicoblastomas (neuroblastomas)

The ganglioneuromas are composed of sympathetic ganglion cells and large numbers of sheathed nerve fibers They arise in the sympathetic ganglia anywhere between the base of the skull and the coccyx and in the medulla of the adrenal the most frequent sites however are at the thoracic levels

Stout states that one fourth of these ganglioneuromas are incompletely differentiated and that these occur in two different forms (1) the ganglion cells of the entire tumor vary in appearance from undifferentiated sympathicoblasts to fully or incompletely differentiated ganglion cells 18 per cent of these metastasize (2) One part of the tumor is a differentiated ganglioneuroma and the other part a sympathicoblastoma (which probably accounts for the reports in the literature of certain sympathicoblastomas spontaneously changing into benign ganglioneuromas) 65 per cent of this type produce metastasis

The sympathicoblastomas (neuroblastomas) are highly malignant tumors of the embryonic sympathicoblast These tumors occur anywhere along the sympathetic chain but the greater majority by far are found in the adrenal medulla They grow rapidly and metastasize freely to bones lymph nodes and liver They commonly contain gross calcium deposits either within a part of the tumor or throughout its entire extent

Both the ganglioneuromas and the sym

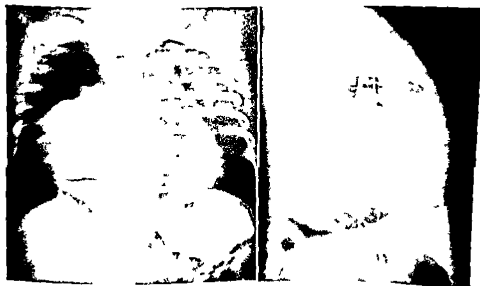


Fig 109 Sympathicoblastoma of the posterior mediastinum in an infant This radiologic examination was made following a diagnostic pneumothorax and proved that the mass was not within the lung Several ribs are eroded in their posterior aspect The esophagus and heart are displaced to the left The transverse line resembling a fluid level in the lower right chest is the shadow of the right diaphragm leaf in its medial portion

pathicoblastomas are found for the most part relatively early in life. Stout states that 60 per cent of ganglioneuromas are found below the age of 20 years. The sympathicoblastomas occur even earlier with the greater preponderance being found in infants and young children. Louria reports that 75 per cent of these are in children less than 10 years of age and that they are usually large in size at their discovery.

Unusual forms of neurogenic tumors sometimes occur. Greenberg has reported a ganglioneuroma in the lower posterior mediastinum that extended along the spinal column through the diaphragm with origins above and below the diaphragm and from both the right and left sympathetic chains.

Tumors of Paraganglionic Cells. These tumors include the benign and malignant pheochromocytomas and the benign and malignant paragangliomas.

Paraganglionic cells are found in both the sympathetic and parasympathetic nervous systems. In the sympathetic nervous system these cells are found in the ganglia extending between the base of the skull and the coccyx and in the medulla of the adrenal. Some of these cells contain chromaffin granules which secrete epinephrine or norepinephrine and these hormonally active tumors are called pheochromocytomas (chromaffinomas); the remaining paraganglionic cells do not contain chromaffin granules and are not hormonally active and tumors arising from these are called paragangliomas.

In the parasympathetic nervous system the paraganglionic cells are found especially in the carotid body, the aortic bodies (also called aortico-pulmonary and cardio-aortic) and certain cranial nerves such as the vagus and the glossopharyngeal nerve and its branches in the middle ear (Stout). As none of these cells contain chromaffin granules they are all paragangliomas.

The greater majority of pheochromocytomas arise in the adrenal medulla but the occasional one is seen in the sympathetic system at the mediastinal level; a case reported by Overholt *et al.* in 1950 was the fourth reported in the literature in this area. Since these tumors produce epinephrine and norepinephrine they usually have an associated paroxysmal hypertension but not always—Maier reported such



Fig. 110. Ganglioneuroma in a 3 year old female. The large mass fills the superior mediastinum. The pulmonary changes are the result of tracheal compression.

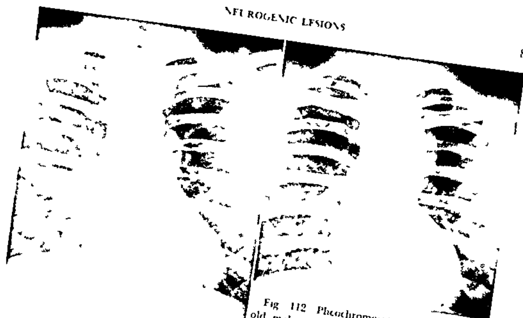


Fig 111 Pheochromocytoma in a 25 year old male. This patient's blood pressure varied between 190 210 mm Hg systolic and 100 120 mm Hg diastolic. Following surgical removal of the lesion the hypertension slowly disappeared over a 2½ year period. Eleven years after operation the patient was still normotensive. From Maier H C: Intrathoracic pheochromocytoma with hypertension. *Ann Surg* 130 1059 1949

Fig 112 Pheochromocytoma in a 16 year old male. This asymptomatic patient had no hypertension preoperatively but during operation his pressure rose to 240 160 mm Hg. After removal of the tumor the blood pressure fell rapidly to 90 70 mm Hg. He completely recovered. From Overholt *et al*: Intrathoracic pheochromocytoma. *Dis Chest* 17 35 1950

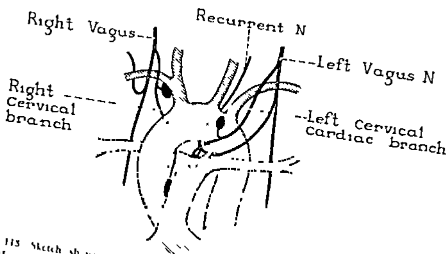


Fig 113 Sketch showing the usual locations of the aortic bodies in the mediastinum. After Monro R S: *Brit J Surg* 38 105 1950



Fig 114 Paraganglioma of the mediastinum in 2 different patients The supradiaphragmatic position of one of these is somewhat unusual From Duncan D K and McDonald J R Chemodectoma (nonchromaffin paraganglioma) of the mediastinum *Am J Clin Path* 21 515 1951

a case of a chemically active mediastinal pheochromocytoma without any definite hypertension preoperatively

Maier also points out that 15 per cent of adrenal pheochromocytomas are bilateral and that occasional cases of coexisting extra and intra adrenal tumors have been reported *he states however that* this has not occurred in the few cases of intrathoracic pheochromocytomas which have been reported

The paragangliomas being hormonally inactive have no associated hypertension the symptoms if any result from the presence of the mass

Paragangliomas arising in the aortic bodies in the mediastinum are rare tumors of special interest these tumors which are also called aortic body tumors and chemodectomas arise from the chemoreceptor (chemodector) tissues and according to Lattes consist of nests and cords of epitheloid cells containing no chromaffin granules and placed in a highly vascular stroma similar tumors occur in the carotid bodies in the cervical area and in the glomus jugularis of the middle ear In general they are benign but at least one in the mediastinum has shown malignant change (Lattes)

Duncan and McDonald state that the chemoreceptor tissue maintains constant physical and chemical conditions in the *blood stream by responding to* (1) decrease in the oxygen tension (2) decrease in the pH of the plasma (3) increase in the temperature of the blood and (4) administration of certain drugs

Grossly these paragangliomas of the aortic bodies are smooth or slightly lobulated well encapsulated and firm and are located in the region of the aortic arch at least one however has been reported in the right posterior mediastinum just above the diaphragm (Duncan and McDonald) Large blood vessels may be present on their surface and this and the vascular stroma may lead one toward a mistaken diagnosis of a blood vascular tumor Usually they can be removed without difficulty

Radiologically most of the intrathoracic tumors of the peripheral nervous system

are located in the posterior mediastinum where they arise from the peripheral nerves near their origin or from the sympathetic and parasympathetic systems these are seen as sharply circumscribed homogeneous masses of varying sizes projecting either to the right or left or in some cases bilaterally Some others such as the paragangliomas of the aortic bodies and an occasional peripheral nerve tumor are not in the posterior mediastinum but rather are further forward in the anterior mediastinum A few tumors extend into the neck from the mediastinum (cervicomediastinal) and at least one has been described which had abdominal and mediastinal components (abdominomediastinal) These extramediastinal components may be apparent on appropriate radiologic examinations

Posteriorly placed neurogenic tumors not uncommonly cause local bone changes this is particularly true of the neurilemmomas neurofibromas Schwannomas and the ganglioneuromas either when circumscribed or when dumbbell shaped with pro



Fig 11b Paraganglioma in a 38 year old female cervicomediastinal in position The mass was palpable in the lower neck The chest examination shows the mass and the extent of displacement of the esophagus and trachea The lesion was surgically removed and weighed 70 grams From McDonald *et al* Chemodectoma (nonchromaffin paraganglioma) of the mediastinum *Ann Surg* *140* 2?1 1954





Fig 11C Erosion of the ninth tenth and eleventh dorsal vertebrae and the right tenth rib by a neurofibroma of the posterior mediastinum Courtesy of Richard A Elmer M D *Atlanta Georgia*



Fig 117 Enlargement of intercostal space and erosion of rib by a posterior neurogenic tumor (ganglioneuroma) Tomogram of the lesion

jections into the intervertebral foramina and spinal canal In Camp s series of neurogenic tumors 65 per cent of the intraspinal types of neurofibromas had bony changes In the ribs the most characteristic of these changes are displacement and thinning of one or more by erosion these findings are usually apparent on conventional chest films In the spine there may be erosion of one or more of the vertebral bodies and pedicles and enlargement of the intervertebral foramina Camp points out that real erosions of the pedicles must be differentiated from pseudonarrowing incident to scoliosis and rotation of the spine Bone changes may not be apparent on conventional views of the chest and may require special techniques such as Bucky films in frontal lateral or oblique positions and tomograms in similar projections

Myelography is useful in demonstrating deformities of the subarachnoid space in cases where tumors arise in the canal or extend into it from the mediastinum Usually these are curvilinear defects which narrow the subarachnoid space to varying extents In cases of neurofibromatosis there may be multiple spinal canal tumors with or without mediastinal components these may lie in the cervical and lumbar areas as well as at the thoracic levels

Pleural fluid accumulations may be seen on chest examinations accompanying both benign and malignant neurogenic tumors the presence of pleural fluid does not jus



Fig 118 Sympathicoblastoma in a 1 year old female who had clinical findings of malnourishment and paralysis of both lower extremities The chest examination discloses a paravertebral mass which is eroding multiple ribs Diagnosis was made by biopsy of a mass presenting in the soft tissues of the back near the mid line Courtesy of Harold O Peterson M D Minneapolis Minnesota



Fig. 119. Bilateral dumb-bell shaped neurofibromata in a patient with neurofibromatosis. The myelogram discloses multiple filling defects in the spinal canal. The mediastinal components of the tumors appear in the apices of the chest.

tify the assumption of malignancy; even a bloody pleural effusion has been reported in association with a benign neurogenic tumor.

Calcium deposits occur, but are uncommon in all types of mediastinal neurogenic tumors except the sympathicoblastoma; in this the picture varies from fine stippling in the center of the tumor to irregular confluent shadows of increased density throughout the mass (Mandeville).

Metastases from malignant growths may be local or distant. Local extensions tend to result in tumors with poorly defined



Fig. 120. Pleural fluid associated with a low grade Schwannoma of the posterior mediastinum. The tumor appears well encapsulated on this chest examination. There is minimal rib erosion. Five years following removal of the mass chest examination showed no recurrence.

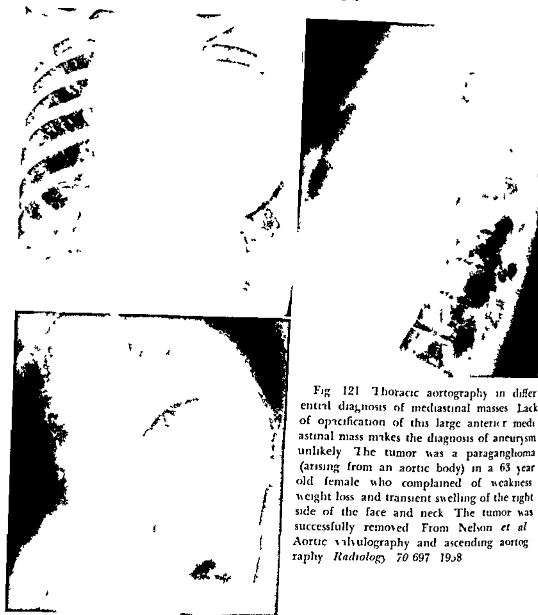


Fig 121 Thoracic aortography in differential diagnosis of mediastinal masses Lack of opacification of this large anterior mediastinal mass makes the diagnosis of aneurysm unlikely The tumor was a paraganglioma (arising from an aortic body) in a 63 year old female who complained of weakness weight loss and transient swelling of the right side of the face and neck The tumor was successfully removed From Nelson *et al* Aortic valvulography and ascending aortography *Radiology* 70 697 1958

borders radiologically caused by the invasion of surrounding structures Distant metastases are most common with the sympathicoblastomas and the structures most frequently involved with these are the bones brain liver and regional lymph nodes

Angiocardiography is helpful in determining the extent of compression and obstruction of the mediastinal vessels by neurogenic tumors and in differentiating those which are adjacent to the larger arteries from aneurysms

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Fig 123 Meningocele of the posterior mediastinum in a 41 year old female with neurofibromatosis Surgical exploration disclosed the cystic mass projecting into the mediastinum between the left sixth and seventh ribs There is erosion of these ribs and of the adjacent vertebrae From Welch *et al* Recklinghausen s neurofibromatosis associated with intrathoracic meningocele Report of a case *New England J Med* *238* 622 April 29 1948

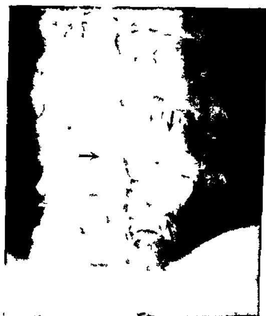


Fig 124 Meningocele of the lower dorsal spine The pedicle adjacent to the lesion is almost completely eroded Surgical exploration disclosed that the cystic mass communicated with the spinal subarachnoid space by a pedicle Courtesy of Robert D Moreton Ft Worth Texas

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MENINGOCELES

Meningoceles are thought to represent an anomalous development of the leptomeninges of the spinal canal resulting in an outpouching of these structures through an intervertebral foramen When such an outpouching occurs at the thoracic level the sac projects into the posterior mediastinum The spinal fluid in the sac communicates with that in the spinal canal through the neck of the meningocele in the intervertebral foramen this opening is frequently quite broad

These sacs may be of any size—so small that they are not apparent on radiologic examination or so large that they protrude widely into the hemithorax They may be single or multiple When single they are usually unilateral

There are two important associated findings with meningoceles The first of these is spinal deformity which occurs at the level of the lesion but occasionally may be at other levels as well The characteristic spinal changes are scoliosis at the level of the lesion erosion of the vertebral bodies and ribs adjacent to the lesion and erosion of the intervertebral foramen through which the lesion originates

A second associated finding is generalized neurofibromatosis which is extremely common in this entity It is far more frequent in this type of neurogenic lesion than in association with solid neurogenic tumors of the mediastinum (Kent reported only 4 patients with neurofibromatosis in 125 patients with neurogenic tumors)

Radiologically, meningoceles may be seen at any level in the posterior mediastinum When single they broaden the mediastinum to one side only Their shape is



Fig 125 Meningocele (a) of the superior mediastinum A diagnostic pneumothorax (b) showed the lesion to be extra pulmonary Surgical exploration confirmed the diagnosis and the lesion was not removed Later contrast media was introduced into the lesion by percutaneous puncture and subsequent radiologic examination revealed the presence of the media in the sac and in the spinal subarachnoid space From Sengpiel *et al* Lateral intrathoracic meningocele *Radiology* 50 515 1948

usually oval or rounded depending upon the resistance to expansion offered by the surrounding structures

Meningoceles are almost invariably associated with spinal deformities There is usually a scoliosis involving several vertebrae There is erosion of the intervertebral foramen pedicle or body of one or more vertebrae depending on the size of the sac and its connection with the spinal canal The adjacent ribs are usually deformed and eroded in a similar manner

The diagnosis can usually be made by



Fig 126 Multiple meningoceles of the mediastinum confirmed by myelography The frontal chest examination discloses a right sided soft tissue mass and rib deformities The vertebral erosions are demonstrated on the lateral tomogram The myelogram discloses the presence of contrast media in bilateral sacs

myelography the communication between the spinal subarachnoid space and the sac is relatively broad allowing free flow of fluid between the two If the sac cannot be opacified in the conventional prone position used in myelography then the spinal needle can be withdrawn from the lumbar region and the patient placed in supine position this allows easier concentration of the contrast material in the kyphotic curve of the dorsal spine where the sac's opening is located

If postoperative films of the chest show evidence of pleural fluid a spinal fluid leak through the area of repair should be suspected



Fig 127 Meningocele of the superior mediastinum in a 31 year old female with neurofibromatosis There are associated deformities of the vertebrae and ribs From Ciaglia P Intrathoracic meningocele *J Thoracic Surg* 23 283 1952



Fig 128 Neurenteric cyst in a 5 year old boy The soft tissue mass widens the right mediastinum and displaces the trachea anteriorly The spinal anomalies are evident there is widening of the neural canal and extensive anterior spina bifida Myelography did not show a communication between the subarachnoid space and the mass but did demonstrate a space occupying lesion in the upper dorsal region anteriorly Laminectomy proved this to be a cystic structure adherent to the cord Thoracotomy three weeks later disclosed that the neck of the mediastinal cyst passed into the bony defect The wall of the excised mass resembled esophagus From Neuhauser *et al* Roentgenographic features of neurenteric cysts *Am J Roentgenol* 79 235 1958

Radiologically mediastinal meningoceles simulate the much more frequently occurring solid neurogenic tumors They should always be suspected in the presence of neurofibromatosis or spinal malformations Myelography is usually required for confirmation of the diagnosis

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NEURENTERIC CYSTS

Neurenteric cysts are rare and unusual lesions of the mediastinum resulting from faulty mesodermal and entodermal development They consist of a cystic mass in the posterior mediastinum connected by a stalk to the spinal cord and meninges through a defect in the vertebral bodies Histologically the wall of the cystic mass resembles a duplication of the alimentary tract The spinal lesions vary but often there are cleft vertebrae

These neurenteric cysts are thought to arise from unobliterated remnants of accessory neurenteric canals of Kovalevski These remnants usually disappear completely during embryonic development but occasionally may persist either as a fibrous cord or as a partially patent tube which may develop into a cyst

The rare lesions of the mediastinum which in the past have been called thoracic diverticula and transdiaphragmatic duplications are probably also neurenteric cysts which have maintained a connection with the alimentary tract In these cases there is a cystic lesion in the posterior mediastinum and posterior extrapleural space usually right sided in position and usually large in size The mass is connected to the duodenum or proximal jejunum in the abdomen by means of a stalk which penetrates the right posterior diaphragm through its own hiatus and joins the mediastinal cyst at its inferior aspect At the superior aspect of the cyst a cord may extend to the deformed spine

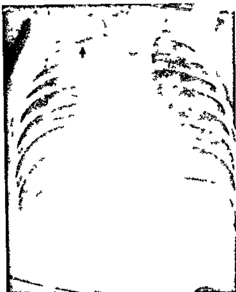
Neuhauser *et al* suggest that these abnormalities may represent only one manifestation of a broad spectrum of anomalies which might conveniently be regarded as resulting from the previous existence of an accessory neurenteric or chorda canal and which may include other such entities as midline dorsal dermoid diastematomyelia duplication of the foregut and anterior spina bifida

Radiologically, neurenteric cysts are suggested by the combined findings of a mediastinal mass and anomalies in the lower cervical and upper dorsal segments of the spine Similar combinations are presented by several other lesions notably mediastinal meningoceles and tumors of peripheral nerves but in these the spinal deformities are likely to be of a different nature (scoliosis erosion destruction)

On radiologic examination neurenteric cysts tend to be rounded oval or lobulated in shape When they communicate with the intestine below there is likely to be some air present in the mediastinal mass

Opaque contrast media can also be useful in identifying those lesions which communicate with the gut The stalk or the mediastinal component or both may be opacified aiding in the diagnosis of the lesion and in the location of its attachment to the intestine Water soluble media are probably preferable to the heavier substances such as barium sulfate and iodized oil

Identification of the type of spinal deformity is extremely important The more common anomalies are a wide neural canal spina bifida and hemivertebra In the most characteristic instances there may be a tunnel extending through the body of



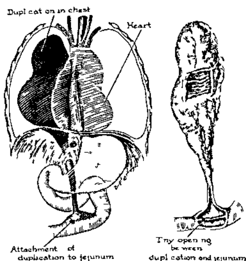


Fig 129 Neurenteric cyst The chest examination discloses a large dumb bell shaped (due to constriction by the azygos vein) mass in the right mediastinum and thorax which contains a small bubble of air at its superior aspect (arrow) The sketch shows the findings at thoracic and abdominal explorations The mediastinal lesion connected with the jejunum in the abdomen by a patent stalk which penetrated the right leaf of the diaphragm There are anomalies of the lower cervical and upper dorsal segments of the spine From Gross *et al* Thoracic diverticula which originate from the intestine *Ann Surg* *131* 363 1950

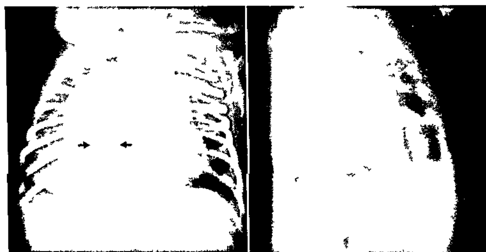


Fig 130 Neurenteric cyst in a 2 month old infant The large fluid filled mediastinal mass contains a small amount of air (arrows) which entered the mass via a patent stalk extending to the small intestine in the abdomen A hemivertebra and other anomalies are present in the upper dorsal spine The cystic lesion was surgically excised Courtesy of John W Hope M D Philadelphia Penna

a vertebra in an anteroposterior direction, through which the remnant of the accessory canal passes Myelography will aid in outlining the spinal canal in the deformed area and in determining any patency of the stalk

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Chapter 14

NORMAL THYMUS. THYMIC TUMORS AND CYSTS

THE THYMUS is an anterior mediastinal gland of unknown function It arises as a paired organ from the epithelial lining of the third pharyngeal pouch During embryonic life it migrates from its cervical position downward into the anterior mediastinum Occasionally there is failure of descent and it remains in the neck Occasionally too a small bit of thymic tissue breaks off and remains in a high position giving rise to ectopic glandular tissue in the neck The lower 2 parathyroid glands also arise from the third pharyngeal pouch and during embryonic life are in intimate relationship with the thymus at times they descend into the mediastinum along with the thymus (see chapter on the parathyroids)

NORMAL THYMUS

The normal thymus is present at birth and continues to increase in size until about 10 years of age During this period however it becomes proportionately smaller in relation to the chest The normal gland varies considerably in size at the same age level It has been shown that thymic tissues are more abundant when the general nutritional state of the patient is at its best At puberty the gland begins to undergo normal involutional changes characterized by a gradual decrease in the number of lymphocytes adipose tissue replacement and atrophy of the epithelial cells

The normal thymus also varies considerably in shape The two lobes may be more or less equal in size or there may be marked asymmetry Caffey states that the transverse diameter of the fetal thymus is greater than the longitudinal and sagittal diameters but that following the onset of respirations the flat thymus is squeezed and then becomes narrower longer and thicker

Microscopically the 2 thymic lobes are divided into lobules by loose connective tissue Each lobule has a peripheral cortical layer composed almost entirely of densely packed lymphocytes (thymocytes) with a scattering of pale reticular cells and a medulla predominating in reticular cells but with a scattering of Hassell s corpuscles Castleman believes that the use of the term thymocyte should be discontinued since most anatomists agree that the small cell of the thymus gland has the same properties as the ordinary lymphocyte

Radiologically the normal thymus during infancy can usually be demonstrated by fluoroscopic examination and on roentgenograms of the chest From birth to about 2 years of age the thymus is relatively large in size in relation to the mediastinum and so is more likely to be identified as a mass during that time After 2 years of age there is more difficulty in visualizing the gland

The identification of the normal thymus is determined by several factors including its size in relation to the mediastinum its shape the stage of respiration the degree of filling of the heart and great vessels and the projections used During a single fluoroscopic examination for example the mediastinum may be enlarged by the gland at certain times and normal at others

Characteristically the normal thymus broadens the anterior and superior mediastinum either bilaterally or unilaterally depending upon the relative size of the 2 lobes The lateral margins of the mediastinum are straight or oval and sharply defined against the lungs Superiorly the gland blends with other structures of the



Fig. 131 Normal thymus in an infant The changing appearance of the thymus with respiration is demonstrated on these frontal chest examinations made in expiration and in inspiration



Fig 132 The normal thymus as it appears on the routine chest examination of 6 different infants

mediastinum or neck and so is not outlined Inferiorly, there is frequently noted on one or both sides a notch between the caudad border of the gland and the structure which it lies against usually the heart This notch may be quite prominent or may be merely a dimple which is seen in certain projections only At times the notch may be more like an angle giving an appearance that has been described as a sail like shadow Whenever possible this notching should be demonstrated fluoroscopically or on films utilizing varied projections as necessary

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THYMIC TUMORS AND CYSTS

Tumors of the thymus occur relatively infrequently they may be benign or malignant Seybold *et al*, in a review of 45 cases of thymic tumors state that the term thymoma should be used to describe only those tumors which are definitely thymic in origin They state It is a slowly growing tumor of the thymus which has arisen from both the epithelial (reticulum) and thymocytic elements of the thymic parenchyma The relative proportion of these cells varies greatly from tumor to tumor and often from place to place in the same tumor Typical Hassall s corpuscles are absent in most thymomas Very common are such features as a dense fibrous capsule distinct fibrous trabeculae the palisading of epithelial cells about cystic

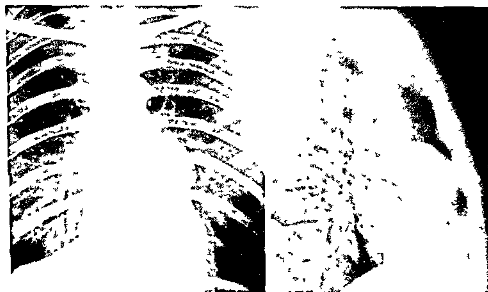


Fig 133 Thymoma in the left anterior mediastinum The egg sized mass rests against the heart base and pulmonary artery With angiocardiographic examination there was no opacification The lesion was easily removed surgically

spaces about blood vessels and about fibrous trabeculae foci of necrosis of cyst formation and of calcification but a number of tumors lack one or more of these features All however contain the thymic lymphocyte or thymocyte and the thymic epithelial cell in varying proportions

The small round cells of the thymus are morphologically identical with lymphocytes and it is difficult to differentiate between thymic tumors predominantly of small round cells and tumors of lymph nodes of the anterior mediastinum In a similar manner other anterior mediastinal masses such as hyperplastic lymph nodes metastatic carcinomas teratomas lipomas fibromas and others may be mistaken for thymomas unless adequate microscopic study is made of sufficient sections of the tumor At times these same lesions and others may arise within the stroma of the thymus but strictly speaking they are not thymomas

Thymomas almost without exception arise in the anterior mediastinum The greater majority are located immediately anterior to the great vessels and the base *of the heart occasionally they are found* at

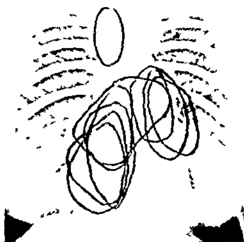


Fig 134 Approximate position and size of 9 thymic tumors in the anterior mediastinum All but one was located in the characteristic position around the base of the heart Courtesy of Brit B Gay Jr M D



Fig 135 Carcinoma of the thymus in a 41 year old doctor who had clinical signs of venous obstruction A frontal roentgenogram discloses minimal widening of the right mediastinal border opposite the aortic knob A venogram shows evidence of obstruction by the mass *Surgical exploration revealed* an inoperable lesion which was subsequently irradiated



Fig 136 Thymic cyst probably congenital in origin This patient had left chest pain and recent hemoptysis Frontal and lateral chest films disclosed a moderately large lesion in the right anterior mediastinum lying against the pericardium At surgery 2 masses were found and removed—one measuring 9 cm and the other 3 cm in diameter Courtesy of Robert D Moreton MD Fort Worth Texas

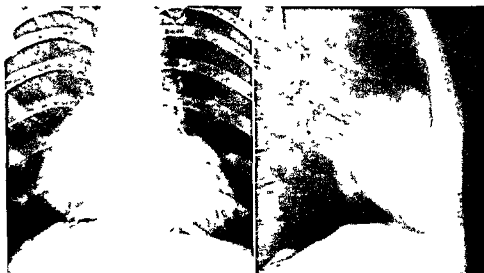


Fig 137 Large malignant thymoma projecting into the right lower mediastinum The small rounded calcific density seen at the lower aspect of the mass in frontal view is actually in the lung posteriorly and not within the tumor

other levels—even as low as the diaphragm. There is a strong tendency for these tumors to invade adjacent structures such as the pericardium, the large arteries and veins, and the adjacent pleura. They rarely spread to distal points.

A thymic tumor occurs in approximately one sixth of patients with myasthenia gravis. In general the thymomas associated with myasthenia gravis are smaller in size than those not associated with this condition. They are usually slow growing and well encapsulated lesions; most are benign, a few malignant. The effect of their removal on the subsequent course of the myasthenia gravis is not uniform.

Of special interest are the rare thymic cysts. These have been described in both infants and adults. Those in infants have been commonly associated with congenital syphilis and have been considered to be secondary to an arrest in the development of the thymus by the infection. Those in adults have been considered by some authors to be true cysts of the thymus; by others to be an extreme degree of cystic change in thymomas. These cysts may be unilocular or multilocular and may contain very little or no recognizable thymic tissue, making accurate identification in some cases difficult.

Radiologically, thymomas are usually discovered on ordinary chest examinations. These masses are almost invariably in the anterior mediastinum and are very frequently located around the base of the heart and the great vessels at their origin. The usual one is well circumscribed, rounded, oval or lobulated in shape, broadens one or both lateral mediastinal borders and displaces the base of the heart and great vessels posteriorly to a variable extent. The larger these masses grow, the more tendency there is for them to spread out laterally and posteriorly, eventually lying to the side of the heart and large vessels as well as in front of them.

Small thymomas may defy detection in frontal roentgenograms of the chest because of the natural width of the mediastinum at the level of the heart base, the shadow at this point being created by the heart and the ascending aorta on the right and the pulmonary artery on the left. But in lateral and oblique studies these lesions can usually be seen and identified as abnormal shadows in the mediastinum.

Thymomas which are invasive may also give some difficulty in identification due to the lack of localized mass formation and the poor definition of the mediastinal borders. In these cases the lesion may be found only through some slight widening of the mediastinal borders or some slight alteration in the normal outline.

Calcifications within the fibrous capsule of the thymus are not infrequent and can be seen as curvilinear lines of increased density in the periphery of the mass. These linear calcifications are not pathognomonic of thymoma as they may be also found in teratomas. Calcification in a thymoma is not a reliable differential point between the benign and malignant lesions.

Angiocardiography is a valuable adjunct when a mediastinal mass is located adjacent to the heart and great vessels in order to differentiate the lesion from an aneurysm. This type examination can also be of help when thymomas invade the vena cava and other large vessels.

The esophagus is usually not deformed by anterior mediastinal masses unless they become very large. But the esophagus may show alterations of its normal motility because of the presence of myasthenia gravis accompanying a thymoma.

Thymic cysts have no distinguishing characteristics which identify them as fluid filled lesions. None that have been reported in the literature were diagnosed as cysts prior to surgery.

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Fig 138 Malignant thymoma The original mass in the anterior mediastinum contained calcium in its wall as shown This lesion was resected but later recurred in the same area causing bilateral enlargement but containing no visible calcium

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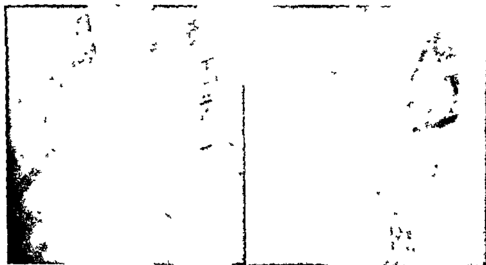


Fig 139 Malignant thymoma lying against the heart and great vessels in the anterior mediastinum These angiocardiograms in oblique projection show filling of the vena cava and aorta but no opacification of the mass

Chapter 15

THYROID LESIONS

THE THYROID is an endocrine gland consisting of a left and right lobe connected by an isthmus. It is situated anteriorly in the lower neck just in front of and to the sides of the upper trachea.

Thyroid enlargements (goiters) may be generalized or localized in type. These lesions can be classified as follows:

(1) Tumor-like lesions—thyroglossal duct cysts, nodular hyperplasia and thyroiditis.

(2) Benign tumors—adenomas of several cellular types (embryonal, fetal, colloid, Hurthle cell, and papillary).

(3) *Malignant tumors—carcinomas of all* types (follicular, papillary, sclerosing, undifferentiated, and Hurthle cell), sarcomas, lymphomas, and teratomas.

Most goiters as they grow remain completely confined to the cervical area. A small percentage, notably discrete adenomas, cysts and multiple colloid adenomatous goiters, extend downward into the superior mediastinum.

Lahey has described the mechanism of this descent as follows: "When an adenoma occurs in the thyroid, particularly in the isthmus or lower pole of the gland, all of *the factors are present for propelling* that tumor into the superior mediastinum. The adenoma rests above an unobstructed superior thoracic strait bounded on the front by *the clavicles and* the sternum and on the sides and back by the vertebrae and the first rib. Any discrete tumor arising in the lower pole of the thyroid gland covered



Fig. 140. Large cervicomediastinal thyroid adenoma presenting mainly to the right. The mass extends well below the level of the aortic arch but is not displacing the arch to any extent. Noteworthy is the separation of trachea and esophagus.

as it is by the sternohyoid sternothyroid and omohyoid muscles which limit upward extension by their attachments to the hyoid and thyroid cartilages and which are inserted into the chest wall in front is subjected to pressure in the downward direction with every act of swallowing

The positions of these goiters in the superior mediastinum are dependent upon their relationships with the anatomical structures in the area Those arising from the isthmus and from the lower poles inferiorly tend to project downward and forward into the anterior part of the superior mediastinum they lie anterior to the recurrent laryngeal nerves the inferior thyroid artery the carotid sheath the innominate and subclavian arteries the innominate veins and on the right the superior vena cava (Sweet) Those arising from the posterior or lateral aspects of the lateral lobes tend to descend into the posterior part of the superior mediastinum behind the structures mentioned above they often rest in a triangular area bounded by the superior vena cava anteriorly the azygos vein inferiorly and the dorsal vertebrae posteriorly



Fig 141 Cervicomediastinal thyroid in a 64 year old female with previous thyrotoxicosis The lower part of the mass lesion extends into the superior mediastinum and displaces the trachea to the right There is associated cardiac enlargement

Goiters are far more common in the right side of the mediastinum than in the left the relatively fixed aortic arch is responsible for this causing deviation of the growing masses to a path of lesser resistance

Practically all benign thyroid masses in the mediastinum maintain some connection with the gland in the neck though this connection is usually broad it may at times be thin and strand like In the occasional case there is no apparent connection between the mass and the gland Wakely and Mulvany explain this as follows As the goiter progresses downward its thyroidal connections become attenuated and no doubt in a few cases completely severed giving rise to the totally intrathoracic type An independent blood supply is however retained in these cases Most investigators are doubtful of the existence of aberrant thyroid tissue even though it is theoretically possible for such tissue to descend into the mediastinum during embryological life Possibly masses can be considered of aberrant origin when they derive their blood supply from the surrounding vessels rather than from above in the cervical area Thyroid masses are occasionally found in the mediastinum as low in position as the diaphragm leaves Malignant thyroid lesions in the mediastinum which have no connection with the gland in the neck are considered to be metastatic in nature

Several terms are used in describing goiters which are partially or wholly within the mediastinum the most common of these are substernal retrosternal partial intrathoracic and complete (or total) intrathoracic It would seem that the terms cervicomediastinal and mediastinal would more accurately describe and locate these masses than the terms now in general use These terms will be used throughout the remaind. this discussion

Cervicomediastinal and mediastinal goiters are quite variable in size—in the series reported by Ellis *et al* they varied from 41 to 1100 grams with an average weight of 218 grams They are both nodular and smooth Their shapes are quite dependent on surrounding structures the cervical component of a cervicomediastinal goiter may be quite obvious on physical examination and its mediastinal extension completely unsuspected unless a roentgenogram is made thyroidectomy in such cases may remove only a portion of the mass

Goiters are usually firm in consistency but may be soft if liquefaction and cyst formation has taken place Calcification is commonly observed in the thyroid mass They may be well encapsulated partially encapsulated or quite invasive Compression of the trachea and the adjacent veins may give rise to alarming symptoms at times Malignant tumors may invade the same structures causing even greater distress Some of the adenomatous goiters found in the mediastinum are toxic but generally not to as great an extent as those confined entirely to the neck Thyroid carcinomas usually have a slow growth adenomas are quite variable in their speed of growth inflammations may be quite rapid

"Plunging" goiters have been described —this is a rare phenomenon in which a cervical goiter becomes temporarily mediastinal in position with coughing

From the radiological standpoint, the importance of chest examination in suspected or confirmed lesions of the cervical thyroid cannot be overstressed The identification and extent of cervicomediastinal and mediastinal lesions can thus be detected

These lesions are predominantly right sided in position due in part to the barrier of the aortic arch on the left Right lobe lesions may be entirely right sided or may have a left sided mass of lesser size Left lobe lesions when small are usually on the left but as they become larger may be diverted to the right by the aortic arch Bilobed lesions and isthmus lesions usually produce bilateral mediastinal widening



Fig 142 Mediastinal metastasis from carcinoma of a cervical thyroid showing growth over a 3 month interval

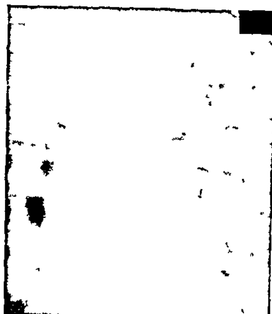


Fig 144 Thyroid adenoma of the left lobe extending as a tongue into the superior mediastinum and displacing the trachea to the right The lesion causes no apparent widening and was discovered at post mortem examination



Fig 145 Large bilobed thyroid adenoma containing extensive deposits of calcium

Less commonly the esophagus is compressed and displaced by goiters in manners similar to the trachea Separation of the esophagus and trachea in the mediastinum is not uncommon The esophageal mucosa is always intact unless there has been invasion of the wall—a sign of malignancy

Both benign and malignant goiters contain calcium (7 in 28 cases reported by McCort 9 in 24 cases reported by Ellis *et al*) and its presence is not helpful in differentiating which type The calcium deposits are usually within the mass but occasionally are at the margins they may be localized or diffusely scattered through the masses The deposits may be conglomerate curvilinear or stippled

Enlarged thyroids commonly compress and displace the adjacent blood vessels particularly the jugular and innominate veins as well as the superior vena cava Angiography is helpful in identifying the extent of involvement Malignant goiters may invade adjacent veins Large goiters extending into the superior mediastinum may also displace the aortic arch downward and to the left in such cases the inferior part



Fig 146 Cervicomediastinal thyroid enlargement compressing and laterally displacing the superior vena cava and several of its tributaries

of the goiter cannot be separated from the aorta fluoroscopically and angiocardiography or aortography may be necessary to differentiate these from aneurysms

Cervicomediastinal and mediastinal thyroids may result in pleural effusion atelectasis of one or more lobes of the lungs paralysis of one or both diaphragms and paralysis of one or both vocal cords these latter 2 findings may be determined by fluoroscopic examination Chylothorax has been reported this was presumably the result of thoracic duct compression

Carcinoma of the thyroid in addition to metastasizing to the mediastinum may also metastasize to other areas such as the lungs (usually slow growing multiple nodular growths) and the bones (usually destructive lesions without surrounding bone reaction in the untreated cases)

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Chapter 16

PARATHYROID TUMORS AND HYPERPLASIAS

THE PARATHYROIDS are endocrine glands which produce parathormone. Normally there are 4 of these glands—2 on the left and 2 on the right; they may vary in number, as many as 7 having been reported. These glands are generally situated just behind the thyroid gland but often the lower 2 are located in the mediastinum.

This mediastinal position of the 2 lower parathyroids can be explained embryologically (Cope). The 2 upper glands have their origin in a primordium arising from the fourth branchial cleft and in their embryologic development remain close to their point of origin; these 2 glands are nearly always located above the level of the lower poles of the thyroid. In contrast, the 2 lower parathyroid glands arise from the third branchial cleft in close proximity to the primordium of the thymus. In their development they travel downward with the thymus as it descends, usually dropping off opposite the lower pole of the thyroid but at times continuing downward into the lower neck or mediastinum along with the thymus. They may even be located in its capsule. The theoretical positions resulting from this abnormal descent are illustrated.

Fig. 147. Lateral and anterior views of the neck and mediastinum showing possible positions (shaded areas) of the lower parathyroids due to differences in development during embryonic life. From Cope, O.: Surgery of hyperparathyroidism: occurrence of parathyroids in anterior mediastinum and division of operation into 2 stages. *Ann. Surg.* 114:706, 1941.

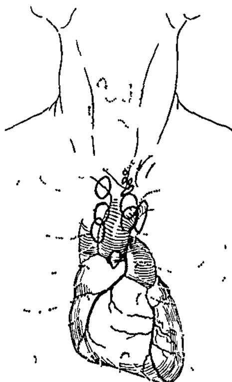


Fig. 148. Parathyroid adenomas of the anterior mediastinum in 11 patients of a series reported by Cope. Diagram showing size and position of the lesions. From Cope, O.: Surgery of hyperparathyroidism: occurrence of parathyroids in anterior mediastinum and division of operation into 2 stages. *Ann. Surg.* 114:706, 1941.

In hyperparathyroidism one or more of the parathyroids enlarge either through development of an adenoma or through

Fig. 149 Parathyroid adenoma (arrows) of the right superior mediastinum proven at post mortem examination. The patient had clinical findings of hyperparathyroidism and signs of skeletal involvement are seen in the clavicles and ribs.

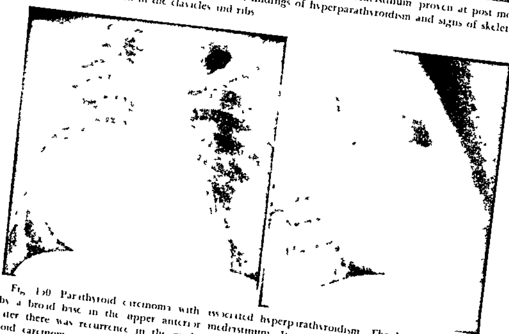


Fig. 150 Parathyroid carcinoma with associated hyperparathyroidism. The huge mass arose by a broad base in the upper anterior mediastinum. It was surgically excised, but 4 years later there was recurrence in the mediastinum. From Weissman *et al.* Mediastinal parathyroid carcinoma with metastasis. *Radiology* 69:352, 1957.

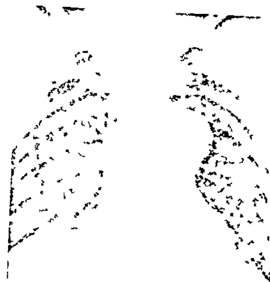


Fig 131 Large parathyroid adenoma of the mediastinum in a 50 year old male with clinical findings of hyperparathyroidism Surgical exploration revealed a mass of 7 cm diameter lying against the anterior side of the main pulmonary artery at its bifurcation From Staub *et al* Mediastinal parathyroid adenoma *Arch Int Med* *85* 765 1950

hyperplasia the former is the more common All of these are functioning tumors in that they produce a hormone similar to if not identical with the hormone produced by the normal parathyroid gland in most instances there is a rough correlation between the size of the tumor or hyperplasia and the degree of hyperfunction

The presence of a parathyroid adenoma or a hyperplastic gland in the mediastinum can be explained in two ways The first is through enlargement of a gland which descended into the mediastinum along with the thymus during embryologic development in these cases the gland is in the superior or anterior mediastinum as would be expected and has a local vascular supply A second method is by gravitation initiated by the weight of the mass and accelerated by the negative intrathoracic pressure these glands are located commonly in the posterior and occasionally in the anterior mediastinum (depending on the relationship of adjacent structures as the gland descends) in these instances the blood supply is derived from the neck

Norris in a study of 322 cases of hyperparathyroidism summarized these facts The exact time of origin of the lesion is difficult to establish Seventy per cent are discovered between 30 and 60 years of age The incidence in females is 3 times greater than in males Multiple adenomas were present in 6 2 per cent of this series The lesions are about equally divided between sides The lower 2 glands are involved far more frequently than the upper ones Ten and seven tenths per cent of these enlarged glands were in aberrant positions (63 3 per cent in the mediastinum 30 per cent in the thyroid and 6 7 per cent behind the esophagus) The smallest adenoma weighed 0 1 grams the largest 120 grams and the average was 12 7 grams The smallest adenoma was more than 10 times the size of a normal parathyroid and the largest was 3 500 times

The parathyroid adenomas are yellow brown in color moderately soft encapsulated and have smooth borders most are ellipsoidal and some are bilobed Cytologically the lesions resemble the normal glands in most cases

Carcinoma of the parathyroids is an extremely rare condition Weissman *et al* have described a case in which a large functioning parathyroid carcinoma was located in the lower right anterior mediastinum Evidently the lesion in the mediastinum was the primary tumor and did not represent a metastasis from a malignancy in the neck

Radiologically, mediastinal parathyroid adenomas and hyperplasias may be identified in 2 ways The first of these is through widening of the mediastinal borders either by virtue of the size of the mass or by its position in the mediastinum As noted earlier they may be found in the superior anterior or posterior parts of the mediastinum In the anterior portion they are not likely to be any further caudad than the

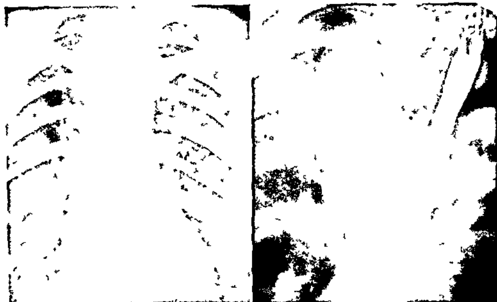


Fig 152 Parathyroid adenoma of the right superior mediastinum The mass broadens the mediastinum to the right and displaces the esophagus and trachea to the left From Wyman S M and Robbins L L Roentgen recognition of parathyroid adenoma *Am J Roentgenol 71* 777 1954

normal position of the thymus gland an exception to this statement however was the large carcinoma mentioned earlier The second method of visualization is by displacement or compression of the trachea or esophagus Wyman and Robbins state that the enlarged glands when in either the neck or mediastinum often produce short smooth indentations in the trachea or esophagus The tracheal compression may be from the lateral or posterior sides the esophageal compression from any side The size of the indentation is roughly related to the size of the mass most being 2 3 cm in length these authors observed compression of the trachea in ⅓ of their 20 cases of the esophagus in ⅓ and of both trachea and esophagus in ⅓

A parathyroid lesion in the neck or mediastinum may be accompanied by radiologically demonstrable lesions elsewhere in the body notably the skeletal system and the urinary tract Adequate investigation of these areas should be made in proven or suspected cases of hyperparathyroidism

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Chapter 17

PERICARDIAL LESIONS

There are 2 types of cysts and diverticula of the pericardium. One type is the congenital pericardial celomic cyst and diverticulum which has its origin during the formation of the pericardial sac. The other is the acquired pericardial inflammatory cyst and diverticulum which results from pericardial inflammation. Primary tumors of the pericardium also occur and are described as mesotheliomas. As these tumors may arise from the pleura as well, they are discussed in a separate chapter. In addition to the primary neoplasms the pericardium is frequently invaded secondarily by various types of adjacent mediastinal masses. Occasionally an anterior mediastinal mass, notably a thymoma or teratoma, will arise within the pericardial sac.

PERICARDIAL CELOMIC CYSTS AND DIVERTICULA

Synonyms for these include simple cyst, springwater cyst, mesothelial cyst and pericardiophrenic angle cyst. The accepted term of celomic cyst was given by Lambert in 1940. These cysts are generally considered to result from failure of fusion of one or more of the mesenchymal lacunae which form the pericardium. The degree of this failure determines whether or not there is a communication with the pericardial sac (i.e., cyst vs. diverticulum).

These cysts and diverticula may occur at any location adjacent to the pericardium but the majority are in the right cardiophrenic angle. They may be attached to the pericardium by either a pedicle or a broad base. The diverticula communicate

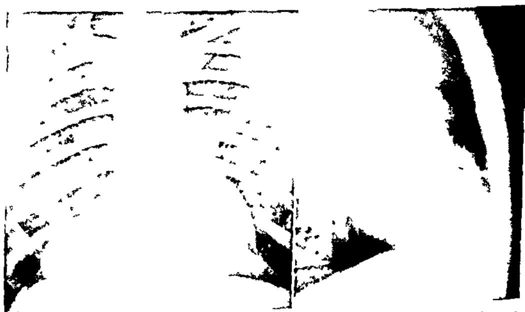


Fig. 153. Pericardial celomic cyst in the characteristic right cardiophrenic angle location. From Fouche, J. W.: Primary intrathoracic nonpulmonary tumors. Am. Surgeon 21:909, 1955.

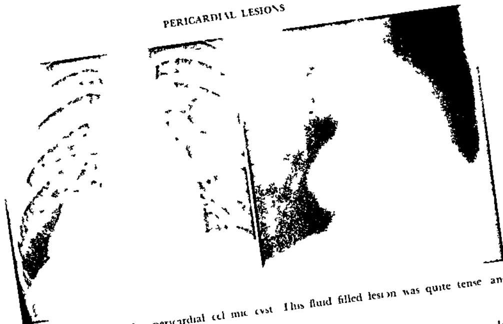


Fig. 154 Multilocular pericardial celomic cyst This fluid filled lesion was quite tense and was easily removed surgically

with the pericardial sac the communication may be small or large in diameter depending on the congenital development Occasionally celomic cysts are located in the superior mediastinum and have no pericardial attachment

Pericardial celomic cysts vary considerably in size the larger reaching the diameter of a grapefruit The diverticula in general are much smaller reaching several centimeters in diameter Most of these cysts and diverticula are unilocular and rounded

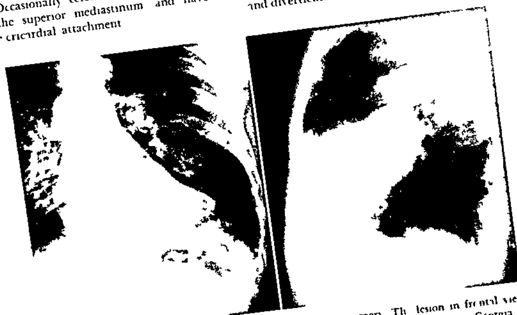


Fig. 155 Left side pericardial celomic cyst confirmed by surgery The lesion in frontal view resembles an epipericardial fat pad Courtesy of Martin Bronson M D Atlanta Georgia

or oval in shape depending on the surrounding structures. Occasionally the cyst may be multiloculated. The fluid within a cyst is translucent and clear; analysis has shown low protein, high chlorides, low sugar and low fat—*similar to a transudate*. Because of the fluid content and the thin wall, the cysts are quite pliable.

Microscopically the wall contains a single layer of mesothelium on a loose fibrous stroma. Inflammation is quite rare and malignant change has not been reported.

Radiologically, these pericardial cysts and diverticula produce rounded or oval shaped bulges on the pericardial outline. In their usual location in the right cardiophrenic angle, they cause an obliteration of the angle in the frontal view and appear as a homogeneous mass resting on the diaphragm anteriorly in the lateral view. The smaller ones tend to extend into the fissure between the middle and lower lobes assuming a "tear-drop" configuration when viewed in the lateral; this "tear-drop" shape is highly characteristic. The larger ones *do not show this sign*.

Fluoroscopic examination may show alterations in shape with deep respiration; this is the result of changing pressures exerted on these pliable lesions by surrounding structures. Pulsations transmitted from the heart are frequently apparent. Alterations in the size of diverticula may be demonstrated when certain positions are assumed by the patient; this is the result of *drainage of the diverticula fluid into the* pericardial sac.

Tomography is frequently helpful in giving a better definition of the lesion, particularly in the frontal projection. A pedicle



Fig 156A



Fig 156B

Fig 156 Pericardial celomic cyst *in the right cardi*ophrenic angle. This small lesion has the characteristic "tear-drop" configuration when viewed in the lateral. The lesion is poorly defined in the frontal view. The sketch shows the findings at surgery. From Rogers, J. V., Jr. and Leigh, T. F.: Differential diagnosis of right cardiophrenic angle lesions. *Radiology 61*: 871, 1953.

hich is not apparent on routine examination may be demonstrated by this procedure

Pericardial celomic cysts in the upper mediastinum will frequently produce smooth walled widening either unilaterally or bilaterally These will resemble other cystic masses occurring at these levels notably those of respiratory and digestive tract origin

These congenital cysts of the pericardium do not calcify In contrast the acquired pericardial cyst may show calcium deposits in its wall

There are 2 other types of lesions which resemble these cysts and diverticula in location and shape when examined radiologically These are the omental hernia tions through the left and right foramina of Morgagni and the epipericardial fat pads on the left and right sides Both of these types are discussed in other sections of this monograph

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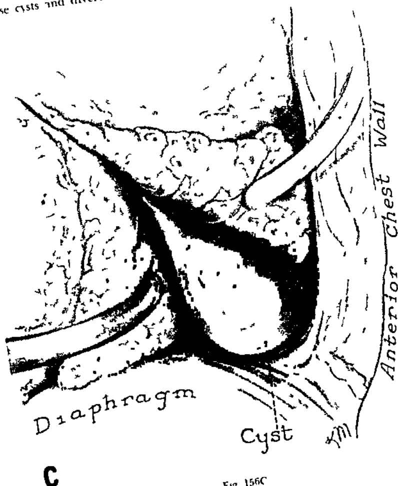


Fig 156C



Fig 157 Pericardial diverticulum This tomographic study of the lesion in the right cardiophrenic angle demonstrates a pedicle which was not apparent on the routine chest examination

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PERICARDIAL INFLAMMATORY CYSTS AND DIVERTICULA

At times cysts or diverticula of the pericardium occur in patients with long standing exudative pericarditis particularly those on a tuberculous basis These lesions have been called by a variety of names including encapsulated pericardial exudates (or effusions) chronic cystic hemorrhagic exudative pericarditis and pseudocyst of the pericardium

Freedman states that these cysts and diverticula occur in patients with adhesive pericarditis when exudate accumulates in a localized pocket of pericardium which has not been obliterated this slow accumulation of fluid causes a progressive enlargement of this localized section of the peri



Fig 158 Celomic cyst of the superior mediastinum The examinations disclose its position against the trachea and esophagus The lesion was easily removed at surgery

cardial sac eventually becoming cyst like in nature

The wall of the pocket may either contain all layers of the pericardium or as sometimes happens the serous layer may herniate through a defect in the fibrous capsule The fluid within the walled-off pocket is usually turbid or serohemorrhagic The heart may be normal in size or enlarged depending on the underlying disease processes

Pericardial calcifications are quite common in these cases the calcium being found not only in the distended sac but in the non distended portions of the pericardium as well

The cysts and diverticula may occur at any point on the pericardium but the greater number of the reported cases have been on the right side anteriorly

On radiologic examination of the chest these lesions are seen as localized bulges in the pericardial silhouette Most of them are broad based smooth or lobulated and sharply defined Some however project outward from the pericardium and communicate with the sac through a pedicle of varying width thus becoming diverticula

These fluid filled cysts and diverticula may show changes in size and shape with respirations elongating in inspiration and becoming shorter and broader in expiration Posture may also have an effect on them as a result of shifting of the contained fluid

Pericardial calcifications are frequent



Fig 159 Right lateral decubitus film showing a pericardial celomic cyst in the right cardiophrenic angle Air injected into the lesion during thoracoscopy confirmed its cystic nature

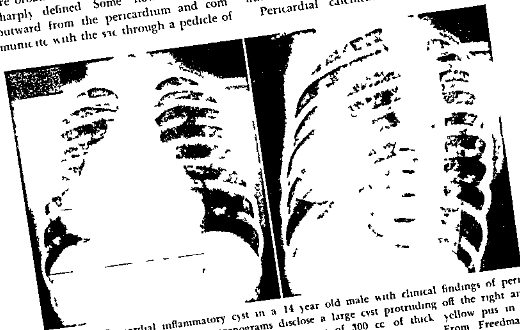


Fig 160 Pericardial inflammatory cyst in a 14 year old male with clinical findings of pericarditis Frontal and oblique roentgenograms disclose a large cyst protruding off the right anterior heart border Operation disclosed the presence of 300 cc of thick yellow pus in a walled-off pocket of pericardium which measured 5 6 mm in thickness From Freedman E. Inflammatory diverticula of the pericardium *Am J Roentgenol* 37 733 1937



Fig 161 Pericardial inflammatory diverticulum in a 48 year old male who had clinical findings of a fluctuating mass to the right of the sternum of 5 years duration A frontal roentgenogram discloses a large bulge on the right heart border A lateral film following aspiration of fluid and instillation of air into the mass on the chest wall confirms a communication with the pericardial cavity From Freedman E Inflammatory diverticula of the pericardium *Am J Roentgenol* 37 733 1937

occurring not only in the distended portions of the damaged pericardium but in the non distended portions as well These plaques can be seen on satisfactorily exposed films of the chest and can be helpful in differentiating this type lesion from others which lie adjacent to the pericardium but do not have associated calcification of the pericardial sac notably pericardial celomic cysts teratomas and thymomas

Transmitted pulsations are frequently observed as would be expected but are difficult to differentiate from intrinsic pulsations Angiocardiography aids in ruling out cardiac and vascular aneurysms or dilatations

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Chapter 18

EPIPERICARDIAL FAT PADS

EPIPERICARDIAL fat pads consist of deposits of mature fat localized around the inferior aspects of the heart on the left and right sides Those on the left usually surround the apex of the left ventricle while those on the right lie against the lateral border of the right atrium The left sided pad occurs with greater frequency than the one on the right but bilateral pads are not uncommon at all If there is a pad on the right there is almost invariably one on the left

Van Fleit has observed that the largest pads tend to occur in young adults He believes that there is no direct correlation between the size of the pads and the general nutritional status of the patient Holt on the other hand comparing chest films made over an interval of time has noted alterations in the size of the pads with alterations in the degree of subcutaneous fat deposits in an individual patient

Grossly these epipericardial fat pads are well defined deposits of adult fat whose surfaces are smooth or mildly lobulated They vary in shape but in general tend towards a pyramidal or oval form They have a good blood supply (Van Fleit)

Their location is invariably in the lower anterior mediastinum on the left and right sides They are bordered medially by pericardium and heart anteriorly by the chest wall inferiorly by the diaphragm laterally and posteriorly by the lung (lingula on the left right middle lobe on the right)

Radiologically epipericardial fat pads tend to have a characteristic appearance In frontal view the pad on the left appears as a smooth or slightly nodular mass around the apex of the heart causing a seemingly enlargement of the apex On the right the pad is seen as a mass resting in the angle between the right atrium and the diaphragm and its lateral border against the lung is straight or mildly curved smooth or slightly nodular

In the lateral projection the left and right pads are partially or completely superimposed They are seen as homogeneous densities in the anterior mediastinum with the anterior surfaces against the chest wall and the inferior surfaces resting

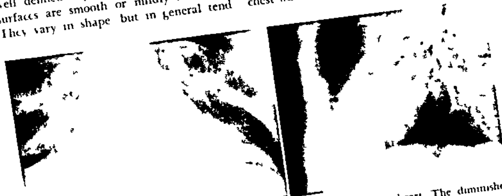


Fig 112 Large epipericardial fat pad surrounding the apex of the heart The diminished density of the triangular shadow suggests the presence of fat tissue



Fig 163 Bilateral epipericardial fat pads

on the diaphragms The borders against the lungs are usually convex and nodular but may be concave and smooth

Since these pads are composed of adult fat they produce shadows of less than water density on the roentgenograms this fact is not too apparent however because of the conflicting shadows of the surrounding structures—the lungs and heart in particular At times the outline of the apex of the heart can be seen through the left sided pad

Tomography particularly in the lateral view frequently gives a clear depiction of the left and right pads and enables one to define their borders more accurately This specialized procedure is particularly useful in differentiating fat pads from several other entities of similar appearance that occur in the cardiophrenic angles notably the pericardial celomic cysts and omental herniations through the foramina of Morgagni

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Chapter 19

MESOTHELIOMAS

In 1942 STOUT and Murray reported their investigations of a group of primary tumors arising from the pleura, pericardium and peritoneum which had similar characteristics. From tissue culture studies it was concluded that these fibrous tissue tumors were of mesothelial origin, although in many respects they resembled the fibromas and fibrosarcomas occurring in other parts of the body. They labeled these tumors mesotheliomas.

Before that time these tumors were known by many names, including endotheliomas, fibromas, fibrosarcomas, sarcomalike tumors, giant sarcomas of the pleura and endothelial sarcomas (Begardus *et al*).

Although mesotheliomas arise from the lining of all 3 serous cavities, the greater majority by far are of pleural origin. The mediastinum is involved when these tumors arise from the parietal pleura lining that area or from the pericardium.

Two forms have been described—the localized and the diffuse. The localized mesotheliomas are well encapsulated masses which are usually lobulated and quite hard. They are attached to the pleura by pedicles which vary in width. If these pedicles are of sufficient length, the tumors may at



Fig. 164. Mesothelioma of the mediastinal pleura in a 54 year old female with chief complaints of general fatigue and weight loss. Chest examination revealed a large mass almost filling the right hemithorax and containing multiple irregular calcium deposits (arrows). The tumor was removed; its gross weight was 1225 grams and its circumference 35 cm. Pathologic diagnosis was mesothelioma of the pleura. From Fouche, J. W.: Primary intrathoracic nonpulmonary tumors. *Am. Surgeon* 21:901, 1955.



Fig 165 Mesothelioma of the pericardium diffuse type in a 27 year old female with clinical findings of congestive heart failure Straw colored fluid was removed from the pleural spaces and grossly bloody fluid from the pericardial space She did not respond and death followed Post mortem examination showed multiple nodulations on the visceral pericardium which microscopically proved to be mesothelioma The pleural fluid may have been on the basis of the accompanying heart failure but could possibly have been derived from the tumor even though there was no apparent involvement of the pleura

times grow in such a way that they are completely surrounded by lung tissue The localized tumors grow to extremely large size In a series of 24 cases reported by Clagett *et al* the diameters varied from 4 to 36 cm In this same series one of the masses weighed 5 000 grams The larger they are the more difficult it is to determine their exact origin

The diffuse mesotheliomas spread along the pleura pericardium or peritoneum and result in marked thickening of these tissues They often invade underlying structures Distant metastases are uncommon

Microscopically the mesotheliomas are divided into two distinct types The fibrous tissue tumors according to Stout and Himadi are composed of spindle shaped cells and connective tissue fibers in a unique arrangement which differs from all other fibrous tumors elsewhere The epithelial tissue tumors are composed of epithelial cells which are capable of forming tubules

Whereas some authors divide mesotheliomas into benign and malignant types others state that all mesotheliomas are malignant

Both the localized and diffuse lesions at times cause effusion This fluid is usually found in the serous cavity that contains the tumor but on occasion there may be fluid elsewhere such as in the opposite pleural space or in the pericardial sac even though no tumor can be identified in these areas

Calcification frequently occurs in the localized types of mesothelioma and is an indication of necrosis within the tumor These deposits are usually confined to one or more small areas of the mass

Radiologically the localized and diffuse forms of mesothelioma present different characteristics The localized form appears as a discrete nodular density frequently lobulated and frequently of large size Only those which arise from the pericardium or from the parietal pleura bordering the mediastinum constitute mediastinal

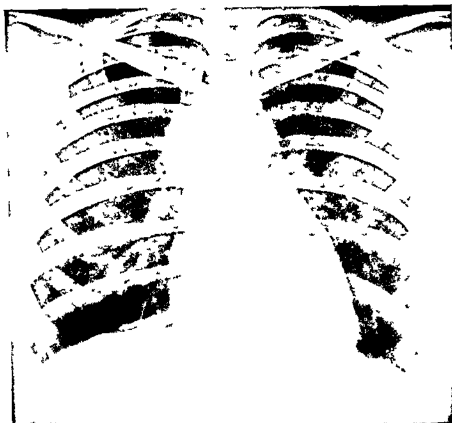


Fig 166A

Fig 166 Localized mesothelioma in a 28 year old female The chest examination (A and B) disclosed a mass in the right anterior mediastinum which was nodular in outline Angiocardiography (C) showed a lateral displacement of the superior vena cava and an unsuspected pericardial effusion (as determined by the position of the opacified right cardiac chambers in relation to the periphery of the pericardial outline) Thoracotomy revealed a 6 cm lobulated mass widely adherent to the superior vena cava the tumor could not be totally excised Seven months later a chest examination (D) revealed recurrence of the tumor pulmonary metastases and pleural effusion From Finby N and Steinberg I Roentgen aspects of pleural mesothelioma *Radiology* 65 169 1955



Fig 166B

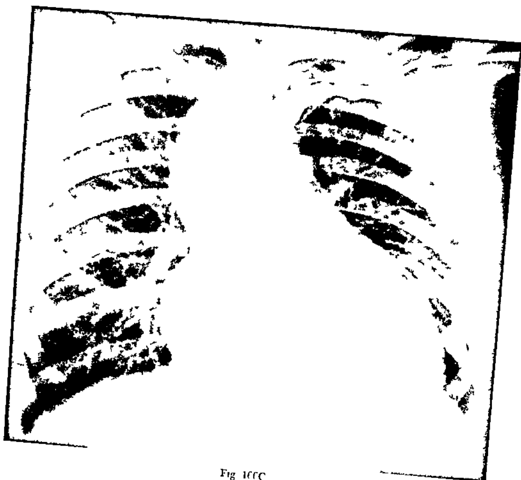


Fig 16C

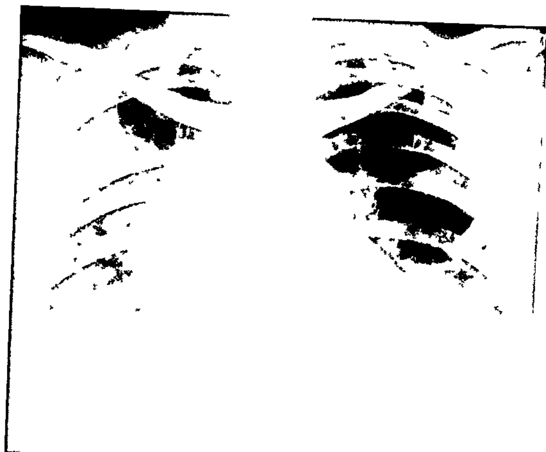


Fig 166D

lesions but those arising from the visceral pleura on the mediastinal side of the lung may appear to be mediastinal masses by the nature of their position In these instances pneumothorax may be necessary to make a differentiation

The calcium deposits contained in localized mesotheliomas which have undergone necrosis are usually seen as punctate areas of increased density confined to one or more small areas of the tumor Over penetrated films are frequently necessary for visualization of these deposits, especially if the lesion is large

The diffuse form of mesothelioma causes nodular thickening of the pleura or pericardial surface On routine examination of the chest this fact may be difficult to evaluate particularly if there is accompanying pleural or pericardial fluid adjacent to the lesion Some help may be gained in the identification of these surface irregularities by making appropriate radiologic examinations following removal of some of the fluid and substitution with air

Angiocardiography is useful in both the localized and diffuse forms of mesotheliomas This procedure aids in the identification of any fluid which may be present in the pericardial sac and in the determination of vascular obstructions and displacements caused by either type lesion

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Chapter 20

BRONCHOGENIC CYSTS

BRONCHOGENIC cysts, along with duplications of the alimentary tract, are congenital lesions which have their origin when the primitive foregut divides into a ventral component to form the respiratory tract and a dorsal component to form the digestive tract. Stout and Lattes state that these various structures lie within an investment of mesenchyme from which develop the fibromuscular tissue of the esophageal wall, the connective tissue, smooth muscle, elastic tissue and cartilage of the bronchi and lungs, the mesothelium of the pleura and all of the mediastinal tissues except the thymus and the nerves, ganglia, paraganglia and chemoreceptor structures *which grow into them from the neural crest*. These authors further state that this complex development accounts for many of the other types of anomalies of the respiratory and digestive tracts, such as tracheo-esophageal fistulae, congenital diverticula and heterotopia. The varied microscopic picture of respiratory and digestive tract cysts is also explained in this way.

In the mediastinum bronchogenic cysts are usually located around the trachea or bronchi but occasionally are found at some distance from these structures. A favorite site is just below the carina. Occasionally one is attached to or is within the wall of the esophagus. Several have been reported as lying in the pericardial sac. One reported case was cervicomediastinal in type and could be displaced entirely into the neck with the Valsalva maneuver.

Grossly, these cysts are rounded or lobulated, being pliable; they tend to mold themselves according to the firmness of the surrounding structures. At times there may be several cysts in a chain. Their outer wall



Fig. 167. Bronchogenic cyst. The paratracheal mass displaces both the esophagus and trachea to the left.



Fig 170 Bronchogenic cyst in a child located between the trachea and the esophagus The lesion is outlined anteriorly by the air of the trachea and posteriorly by barium in the esophagus Courtesy of Richard A Elmer M D Atlanta Georgia

is smooth but the inner wall is frequently trabeculated Usually they consist of a single cavity but ocasionally one may be multiloculated or contain several non communicating cavities of varying sizes The non infected cysts contain thick mucoid material which is white or dirty brown the infected ones contain purulent material

Microscopically their walls contain all or most of the elements seen in the normal bronchus but in different proportions these include mucous glands fibrous tissue elastic tissue smooth muscle fibers cartilage and an interlining of ciliated columnar epithelium Infection may destroy the wall Apparently they do not calcify Malignant degeneration (sarcoma and carcinoma) has been reported in pulmonary cysts but none in those of the mediastinum

The cyst may be firmly adherent to the trachea or bronchus or loosely attached



Fig 171 Bronchogenic cyst Surgical exploration disclosed that it was in the mediastinum and communicated congenitally with a bronchus of the right lung An air fluid level is seen on the films made with the patient in upright position The cyst was infected—a common finding when there is a communication with an air passage (trachea or bronchus) From Brown R K and Robbins L L The diagnosis and treatment of bronchogenic cysts of the mediastinum and lung *J Thoracic Surg* 13 84 1944

by areolar tissue At times there may be a pedicle Occasionally one communicates with the trachea or a bronchus in which case it is very prone to infection

Radiologically, bronchogenic cysts are seen on routine chest examinations as smoothly outlined masses within the mediastinum broadening this area unilaterally or bilaterally Characteristically they are located around the trachea and may deform this structure to some extent If attached to the trachea they will ascend with swallowing If between the trachea and the esophagus they tend to separate and compress these structures this is best demonstrated in the lateral projection and with barium swallow If the cyst is in the subcarinal area it may elevate and flatten the bronchial angle and this can be demonstrated on films which are sufficiently penetrated to show the air columns in the bronchi Occasionally one of these may be located low in the mediastinum and present as a rounded or oval shaped mass far removed from the trachea or bronchi

When there is a congenital communication with the trachea or one of the bronchi air and fluid will frequently be present and a fluid level can be demonstrated if the patient is examined in an upright position The exact point of communication may be visible on routine films fluoroscopy or tomograms but more likely an opaque contrast media will be necessary to demonstrate the tract

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Chapter 21

DUPLICATIONS OF THE ALIMENTARY TRACT

DUPLICATIONS of the alimentary tract are known by several names including esophageal cyst gastric cyst enteric cyst enterogenous cyst inclusion cyst para esophageal cyst archenteric cyst and fore gut cyst The term duplication of the alimentary tract was suggested by Ladd many years ago

The origin of these cysts from the primitive digestive tract was described in the preceding chapter They are found not only in the mediastinum but at any point along the digestive tract from the mouth to the anus In the mediastinum they are usually in the middle third posteriorly adherent to the esophagus though there is occasionally no attachment They vary greatly in size and shape

Microscopically the duplication s wall contains mucosa submucosa and muscularis The mucosa is epithelial in origin and resembles that of the esophagus stomach or intestine usually there is a mixture of these types in the same cyst Occasionally salivary tissues are present The muscle coat is in intimate relationship to the adjacent esophagus and may be indistinguishable from it When gastric mucosa is present an ulcer may form in a manner similar to the peptic ulcer of the stomach Perforation of the ulcer may result in a fistula with the esophagus trachea bronchus or some other adjacent structure

The cystic fluid is as a rule is clear colorless and of mucoid consistency With necrosis in the wall the fluid is cloudy or bloody When gastric mucosa is present the fluid is acid When the secretory mucosa is still active the fluid continues to accumulate enlarging the cyst

These cysts are often symptomatic and

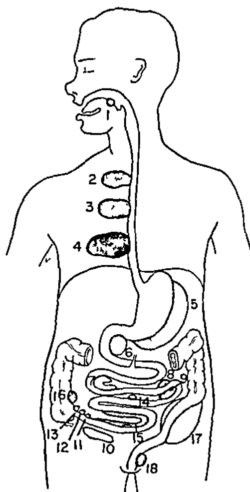


Fig 172 Sketch showing the distribution of duplications in 18 cases Three of these are in the mediastinum From Ladd W E and Gross R E Surgical treatment of duplications of the alimentary tract *Surg Gynec & Obst* *70* 295 1940

because of this are frequently discovered in infancy or childhood The symptoms result from (1) pressure on adjacent structures notably the esophagus trachea and

bronchi (2) functional activity (3) infection

Radiologically, these duplications of the alimentary tract are seen as mediastinal masses of varying size usually located against the esophagus in its middle third They more frequently bulge to the right than to the left but at times may extend bilaterally

Esophageal studies commonly show a smooth curvilinear defect with tapering upper and lower borders These defects caused by the cysts are frequently similar to those produced by benign tumors of the esophagus for a description of these findings the reader is referred to the section on tumors of the esophagus

Opacification of the cyst during an esophageal study indicates a fistulous communication between the 2 this usually is the result of erosion and perforation

Rarely a cyst may erode one or more ribs and the dorsal spine For this reason a study of the spine in frontal lateral and oblique projections is indicated When erosion is present there may be difficulty in differentiating between duplication and neurogenic tumor

Neurenteric cysts which are closely akin to duplications of the alimentary tract also have associated spinal deformities but these are of a congenital nature and thus different from those seen with ordinary duplications of the alimentary tract These cysts are discussed elsewhere in this text

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Fig 173 Duplication of the alimentary tract in an infant The lesion is located in the posterior mediastinum and projects forward and to the right The esophagus and other mediastinal structures are displaced to the left The mass was successfully removed Courtesy of John W Hope M D Philadelphia Penna

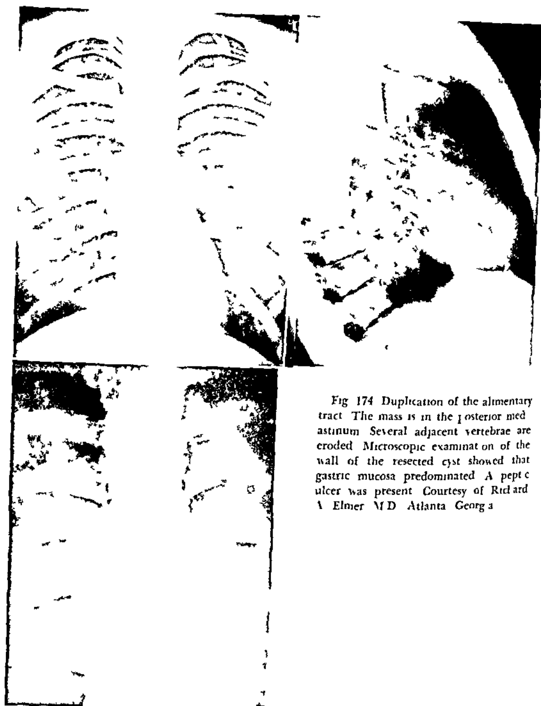


Fig 174 Duplication of the alimentary tract The mass is in the posterior mediastinum Several adjacent vertebrae are eroded Microscopic examination of the wall of the resected cyst showed that gastric mucosa predominated A peptic ulcer was present Courtesy of Richard A Elmer M D Atlanta Georgia

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Fig 175 Duplication of the alimentary tract *paraesophageal in position The rounded mass* compresses and narrows the esophageal lumen Courtesy of Richard A Elmer M D Atlanta Georgia

Chapter 22

TUMORS OF THE TRACHEA

TUMORS ARE the predominant mass forming lesions of the trachea These may be either primary or secondary The primary growths are relatively rare, these will be discussed in this chapter The secondary growths are frequent, and are due either to direct extensions from neighboring tumors in the mediastinum, or to metastases from more distant growths

Many different types of primary tracheal tumors have been reported, including carcinomas, sarcomas, cylindromas, mixed tumors, angiomas, endotheliomas, papillomas, fibromas, lipomas, leiomyomas chondromas, osteomas, adenomas, intratracheal goiters, and amyloid tumors These occur in both adults and children, one large reported series revealed that of 503 tumors in adults 247 were malignant, and 256 benign, whereas of 43 tumors in children 3 were malignant and 40 benign All malignant tumors in the children were sarcomas, and 78 per cent of the tumors in adults were carcinomas

The largest and most important group of primary tumors are the carcinomas In one reported series of 433 primary tumors of the trachea, 147 were of this nature These tumors may be squamous cell cancers, adenocarcinomas, or a special form of adenocarcinoma known as cylindromas The latter have a distinct biologic behavior different from other adenocarcinomas, namely, a slow rate of growth a high rate of invasiveness and recurrence after removal, and a definite potentiality for metastasis They are the second most common malignant tumors of the trachea

The squamous carcinomas arise from the squamous cells of the tracheal wall, and are either fungating or ulcerating in type, one interesting aspect of these tumors is that they have apparently not increased in incidence though similar tumors of the bronchi have done so The adenocarcinomas originate from the mucosa of the trachea and grow as polypoid or ulcerating lesions Both of these types of carcinomas arise most commonly on the lateral and posterior walls of the trachea in its lower third They tend to grow inwardly with encroachment on the tracheal lumen causing infrequent tumefaction of the mediastinum Early symptoms and signs due to partial tracheal obstruction are the rule Those located near the carina may encroach on the bronchi, and at times, cause atelectasis or emphysema Metastatic spread to the mediastinal lymph nodes or adjacent organs may occur

The cylindromas probably arise from the cells lining the mucous glands of the trachea rather than from the surface epithelium They are histologically similar to the tumors of the same name found in the salivary glands and elsewhere The mucosa over them is intact, and although they are likely to encroach on the tracheal lumen to some extent, their predominant growth is usually outward with resultant mediastinal tumefaction They usually arise in the upper third of the trachea, and as mentioned previously, are slow in growth and tend to infiltrate and recur after removal

At times, primary carcinomas of respiratory tract origin are found in the mediastinum with no obvious attachment to the trachea or the proximal bronchi These are thought to arise from tissues which were split off from the primitive foregut at the time of its division into the respiratory and digestive tracts Conceivably some of those

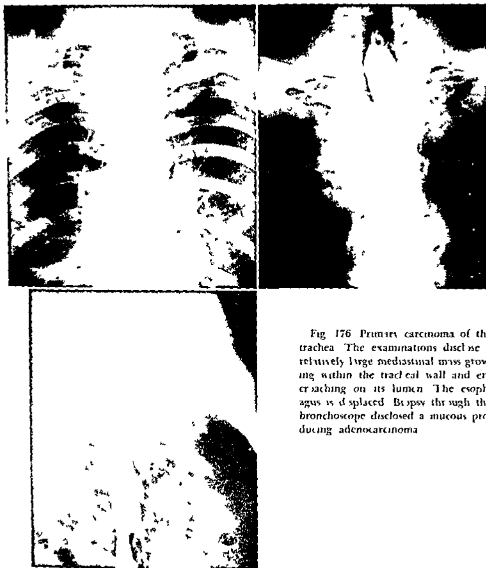


Fig 176 Primary carcinoma of the trachea The examinations disclose a relatively large mediastinal mass growing within the tracheal wall and encroaching on its lumen The esophagus is displaced Biopsy through the bronchoscope disclosed a mucous producing adenocarcinoma



Fig 177 Primary carcinoma (cylindromatous type) in a 47 year old male The tumor is located in the middle third of the trachea and is predominantly intraluminal This radiologic examination was made with a 250 KVP therapy machine and localized the lesion in relation to the radiation field Courtesy of Harold W Jacox MD New York N Y

attached to the trachea may be of similar origin

Benign tumors of certain types may grow peripherally from the tracheal wall to produce obvious mediastinal lesions but the greater majority arise intraluminally Many of them are pedunculated and may move upward and downward in the tracheal lumen intermittently blocking the air passages

Radiologically, a primary tumor of the trachea may be recognized as a mass in the mediastinum as a bulge into the tracheal air column or as a combination of both Frontal lateral and oblique projections are frequently necessary to determine the relationship of the mediastinal tumor to the trachea For the demonstration of intraluminal lesions special techniques are frequently necessary fluoroscopic spot films tomograms and studies using contrast media have proven to be helpful One projection may demonstrate the lesion better than another depending upon the position of the tumor It should be remembered that most of these are located on the posterior or lateral walls and that they are rarely encircling in type

Pedunculated growths in the trachea may change in position with respiration or with changes in the position of the body In these cases fluoroscopic observation particularly with image amplification is the method of choice

The upper trachea is usually well seen in frontal and oblique projections but is partially or totally obscured in the lateral view when the patient is positioned in the conventional way with the arms above the head In such instances the trachea may be rendered visible in the lateral view if the hands are clasped tightly in behind and the shoulders are rolled backward This maneuver is more easily accomplished in the thin individual than in the obese

Nontracheal growths in the mediastinum which encroach on the trachea and cause compression and displacement of this structure may strongly resemble primary growths and a differentiation between the two may not be possible radiologically

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Chapter 23

ESOPHAGEAL LESIONS

SEVERAL esophageal lesions produce significant mass formations in the mediastinum. The most common of these are the benign and malignant tumors, megaesophagus and epiphrenic diverticula. Pharyngeal diverticula strictly speaking are not primary esophageal lesions, but they lie in such intimate contact with the esophagus in the lower neck and upper mediastinum and cause such marked compression and displacement of it that they are included in this section for discussion.

Other lesions of the esophagus do not usually produce significant mediastinal shadows. Rarely traction diverticula and esophageal varices may attain sufficient size to be visible on chest examinations. Several lesions indirectly related to the esophagus are discussed elsewhere in this monograph; these include duplications of the alimentary tract, mediastinal abscesses resulting from esophageal perforations and esophageal hiatus hernias.

PHARYNGEAL DIVERTICULA

Pharyngeal diverticula are pulsion diverticula arising as mucosal outpouchings between the oblique fibers of the inferior constrictor muscle and the circular fibers of the cricopharyngeus muscle of the hypopharynx. These diverticula are known by several other names including pharyngoesophageal pouches, Zenker's diverticula, pulsion diverticula of the upper esophagus, retrocervical diverticula and deep pharyngeal pressure diverticula. The rarity of this type of diverticula in children indicates that they are of an acquired nature.

At the juncture of the oblique and circular muscle fibers of the hypopharynx there is a weak point in the posterior wall. Frequently there is a mucosal dimple anatomically at this point. But occasionally an outpouching of mucosa develops where these two sets of fibers meet. Negus believes that there are several factors in the development of pulsion diverticula of this nature; he lists them as follows: (1) the low position of the larynx and with it the descent of the mouth of the esophagus, (2) the obliquity of the inferior constrictor muscle in relation to the circular arrangement of the cricopharyngeus, (3) the attachments of the anterior wall only of the esophagus to the larynx and its wide range of motion during swallowing, (4) the active contractions of the cricopharyngeus sphincter and the difficulties of coordination of its relaxation with contraction of the pharyngeal force pump, (5) the lack of longitudinal suspension of the posterior wall of the mouth of the esophagus and (6) the absence of support at the triangular area in the posterior wall.

Pharyngeal diverticula may be quite small in size and remain so during life. The tendency is, however, for them to enlarge with the passage of time. As they grow they project inferiorly (because of rigid enclosure on all other sides) down the lower neck and into the superior mediastinum. With increasing size the mouth of the sac comes more into line with the pharyngeal lumen; then the esophageal opening appears as a narrow aperture on the anterior border of the neck of the sac. In such instances ingested material is likely to fill the sac immediately on swallowing. Compression of the upper esophagus is a constant finding, and the degree present is dependent upon the size of the sac and the amount of material contained within its lumen.

The wall of the diverticu



Fig 177 Primary carcinoma (cylindromatous type) in a 47 year old male The tumor is located in the middle third of the trachea and is predominantly intraluminal This radiologic examination was made with a 250 KVP therapy machine and localized the lesion in relation to the radiation field Courtesy of Harold W Jacox M D New York N Y

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pharynx and esophagus with contrast media Small diverticula may fill and empty quite rapidly not retaining the media for any considerable period of time In these cases study in the oblique projections is indicated since in the frontal projections the opacified esophagus may completely obscure the small lesion Larger pharyngeal diverticula practically always fill with the initial ingestion of contrast media this is because of the relationship of the sac to the pharyngeal lumen Again the oblique projections are the most satisfactory for examination for in these positions not only can the sac itself be investigated but the equally important esophageal deformity can be seen

The fact that these diverticula so frequently resemble other superior mediastinal masses on routine chest examinations but yet are so quickly diagnosed by contrast media studies emphasizes the need for such studies in every single case of mediastinal mass formation

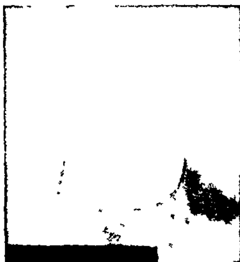


Fig 180 Pharyngeal diverticulum This lateral projection shows the barium filled diverticulum between the esophagus anteriorly and the cervical spine posteriorly The esophageal compression and deformity is quite characteristic

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EPIPHRENIC DIVERTICULA

Epiphrenic diverticula are sac like outpouchings of the lower esophagus resulting from herniations of mucosa through the muscle layers of the esophageal wall Synonyms for these diverticula include supradiaphragmatic diverticula intrapleural diverticula and esophagoceles

These are pulsion diverticula and are similar to the pulsion diverticula of the lower pharynx They usually develop on the right posterior wall of the esophagus about 5 to 10 centimeters above the cardia and project to the right although their etiology is indefinite their frequent occurrence at this point suggested to Harrington a possible embryonic origin due to a congenital defect in the wall of the esophagus

When they are small they represent a bud like outpouching of the esophageal lumen with the stalk and sac of the diverticulum being in the same transverse plane But as they grow larger and as the sac and its contents become heavier they often gravitate to a more dependent position below the level of the stalk's esophageal opening this pendulant sac not only causes compression of the lower esophagus but also alters the relationship of the esophagus and diverticulum in such a way that the pouch becomes a continuation of the esophageal lumen ingested material may first enter the sac and then spill over into the distal esophagus

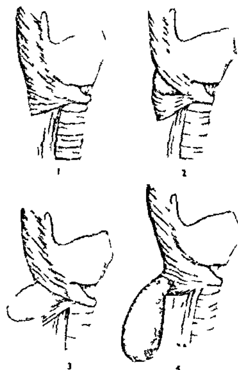


Fig 178 Pharyngeal diverticulum These four sketches demonstrate its location and its mode of development between the circular and oblique fibers of the pharyngeal musculature From Negus V E Pharyngeal diverticula *Brit J Surg* 38 129 1950

mucosa and submucosa Occasionally some fibers from the oblique and circular muscles are incorporated in the sac s outer wall

At least two unusual forms of these diverticula have been described One type *dissected downward between the mucosa* and muscularis of the esophagus thus becoming an intramural diverticulum rather than the usual extramural type The other arose laterally rather than posteriorly at a point in the musculature where normally passes the recurrent laryngeal nerve a branch of the inferior thyroid artery and vein and a bundle of lymphatic vessels

Radiologically, pharyngeal diverticula can be *definitely identified and diagnosed* Occasionally the larger ones are initially discovered on routine chest examinations where they are seen as masses in the superior mediastinum projecting bilaterally or unilaterally (the left side predominating) and having smooth walls They may appear homogeneous or mottled depending on the amounts of air and fluid present and in *the upright position they may show an air fluid level*

More commonly however these diverticula are first demonstrated by studies of the

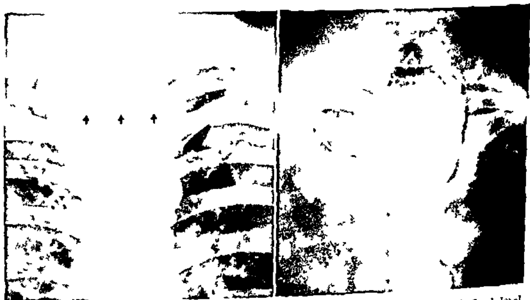


Fig 179 Pharyngeal diverticulum projecting into the superior mediastinum A fluid level is apparent in the chest examination made in upright position Esophageal studies with contrast media confirm the diagnosis

Histologically, the wall of this type diverticulum consists of a mucosa, submucosa, muscularis mucosa and a fibrous layer of varying density Chronic inflammation of the wall is very common, and is due to stasis of contents which have been ingested or regurgitated Ulcerations are not uncommon, and may be the source of such clinical findings as pain, hemorrhage, perforation and obstruction

Chronic inflammation is common in the short segment of the esophagus distal to the diverticulum's opening and may result in spasm or stricture Some authors believe that in some cases this spasm represents true cardiospasm and that the resultant *increased intraluminal pressure initiated* the formation of the diverticulum

Epiphrenic diverticula are not to be confused with duplications of the alimentary tract, which occur in the mediastinum, and which are usually adjacent to the esophagus The occasional one of these located in the lower mediastinum may perforate *into the esophagus as a result of ulceration* or other cause, and may resemble a diverticulum These lesions are discussed in another section of the book

Radiologically, epiphrenic diverticula are frequently first discovered on routine chest examinations, where they are seen as masses in the mediastinum just above the diaphragms, frequently having air fluid levels On these films they resemble hiatal hernias in most respects, except that they *are more likely to be asymmetrical in relation to the midline than are hiatal* hernias



Fig 183 Epiphrenic diverticulum in a 52 year old male An oblique view discloses the characteristic position of the lesion on the posterior side of the esophagus The distal esophagus appears to be spastic possibly as the result of esophagitis a frequent associated finding Microscopic examination of the resected diverticulum disclosed considerable chronic inflammation in its wall Courtesy of J H Bowen M D Maryville Tennessee



Fig 184 Epiphrenic diverticulum in an asymptomatic 42 year old patient The lesion filled promptly with the first swallow of barium The distal esophagus is not significantly deformed

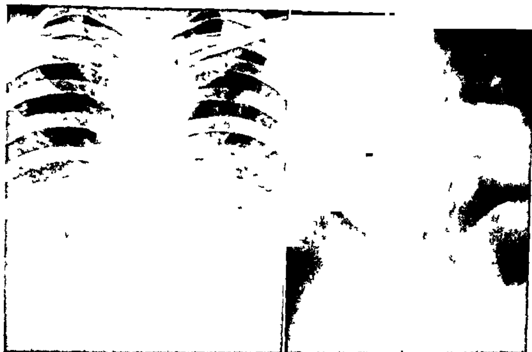


Fig 181 Epiphrenic diverticulum The chest examination discloses a mass just above the right diaphragm leaf medially which has an air fluid level An esophageal study reveals the true nature of the lesion The barium column in the esophagus is continuous with that in the diverticulum The esophagus distal to the lesion is collapsed



Fig 182 Epiphrenic diverticulum showing a small fluid level (arrow) in the lateral view of the chest The lesion is confirmed by esophageal studies

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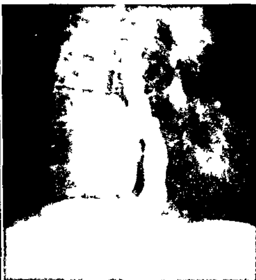


Fig 184 Epiphrenic diverticulum in an asymptomatic 42 year old patient The lesion filled *promptly with the first swallow of barium* The distal esophagus is not significantly deformed

Their true nature can be identified by contrast media studies of the esophagus. Characteristically, the upper two thirds of the esophagus is normal or slightly widened in diameter. The sac lies to the right and posteriorly and its opening into the esophagus is 5 to 10 centimeters above the level of the cardia. Ingested media fills the sac immediately. The segment of esophagus distal to the sac's opening is frequently narrowed and the media often traverses it in spurts rather than in a continuous stream. This narrowing is the result of spasm, stricture, compression or combinations of these. The esophagus shows no redundancy. The entire stomach can be identified below the diaphragm unless there is a hiatal hernia associated with the diverticulum (not an uncommon occurrence). In these cases 2 separate masses will be seen in the mediastinum—one representing the diverticulum, the other being the hernial sac. The relationships of the lower esophagus to the diverticulum and the hernia will depend on the type of hernia present and the position of the two masse

The diverticulum is likely to retain som of the contrast media continuously throug out an examination. Regurgitation of th sac's contents into the esophagus is likel to occur when the patient is in recumber or Trendelenburg positions.

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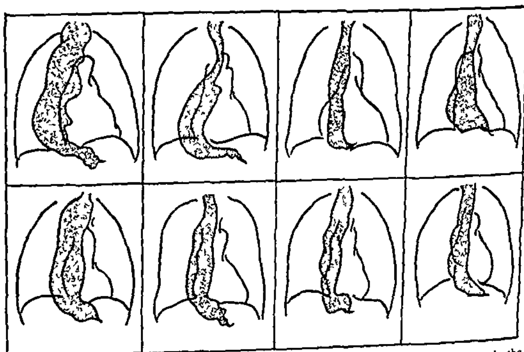


Fig. 185. Diagrammatic representation of megaesophagus and its relationships with the heart. From Weens, H. S.: Pulmonary disease associated with megaesophagus. *Am. J. Roentgenol.*, 52:472, 1944.

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MEGAESOPHAGUS

Megaesophagus is defined as a generalized dilatation of the esophagus the cause of which is not fully understood In some instances a congenital absence of the ganglionic cells from the myenteric plexus has been found resulting in failure of proper relaxation of the lower esophageal segment

The enlarged esophagus usually projects *anteriorly and to the right the spine and* the aorta tend to prevent posterior and left sided extension The esophagus may be so large that it extends to the right lateral costal wall In long continued cases having marked intraluminal pressure the esophageal wall is thinned and the musculature stretched Retention of solids liquids and air within the lumen is common and aspiration of these contents frequently results in pulmonary suppuration

Radiologically, the dilated esophagus appears opaque when filled with retained fluid mottled with fluid and air translucent with air only Technical factors and projection largely determine how it appears in a given situation A fluid level may be present in the upright position A double air shadow is sometimes seen in the frontal projection and is the result of the trachea superimposed on the esophagus

A diagnosis of megaesophagus is confirmed by barium studies which will show enlargement of the esophageal lumen and stricture at its lower end

The pulmonary changes which frequently accompany megaesophagus may be seen on appropriate films of the chest these include such lesions as abscess bronchiectasis, acute and chronic pneumonitis (including lipoid pneumonia) and chronic fibrosis Some of these may be suspected on routine examination while others may require additional studies such as bronchography

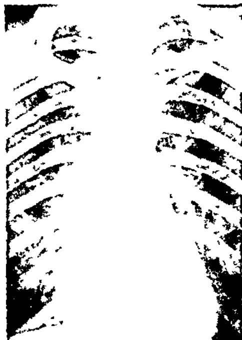


Fig 186 Megaesophagus The lower right border of the esophagus simulates the border of the right atrium and is indistinguishable from it in this view The esophageal lumen contains fluid

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TUMORS OF THE ESOPHAGUS

For purposes of discussion the tumors of the esophagus can be divided into the benign and malignant types

The benign tumors are quite varied in type and include such entities as the leiomyomas polyps lipomas fibromas vascular tumors lymph tumors chondromas neurofibromas and others The most common of these are the leiomyomas which are smooth muscle tumors arising in the wall of the esophagus these are extramucosal

A

B

C

D

Fig 187 Leiomyoma of the esophagus The frontal chest film (A) discloses the mass in the upper right mediastinum With barium studies (B C and D) the esophageal deformity varies with the degree of filling and with the projections used

mural lesions which grow inwardly encroaching on the lumen and outwardly producing mass formation. They do not invade the mucosa and may be single or multiple. They usually occur in the middle or lower third of the esophagus and in their growth tend to expand the mediastinum to one or the other side or bilaterally depending on size and position. The tumors may be quite large when initially discovered; one weighing 900 grams has been reported.

The primary malignant tumors of the esophagus include the carcinomas, the sarcomas and the carcinosarcomas. The greater majority of these by far are the squamous cell carcinomas which arise in the covering squamous mucosa and grow in all directions. The second most common types are the adenocarcinomas which usually arise in the glands of the stomach at the cardia and invade the esophagus secondarily, but in some cases probably arise from slightly displaced cardial glands located a few millimeters within the lower esophagus under its squamous epithelium; in these instances they can be classified as primary tumors by a narrow margin. The sarcomas and the carcinosarcomas are quite rare; one interesting characteristic of these is that they are frequently pedunculated (Stout and Lattes).

A third group of mass forming lesions of the esophagus are those of congenital origin; these include congenital cysts, cystic duplications and heterotopic epithelium in the esophagus. The congenital cysts and duplications which presumably have their beginning when the foregut splits into its ventral and dorsal components to form the respiratory and digestive tracts are discussed in a separate section of this book.

Radiologically, benign and malignant tumors of the mediastinal esophagus are frequently discernible as soft tissue masses on routine chest examinations. The posteromedial borders of the left and right lungs closely approximate the esophagus throughout its entire length in the thorax and expanding lesions of the esophagus tend to deform these borders. Large esophageal tumors present no problem in their identi-



Fig. 188. Leiomyoma of the lower esophagus shown as a retrocardiac oval shaped mass on frontal and lateral roentgenographic studies.



Fig 189 Unusual leiomyoma of the lower esophagus and upper stomach (A D) From Harrington S W Surgical treatment of benign and secondarily malignant tumors of the esophagus *Arch Surg* 58 646 1949

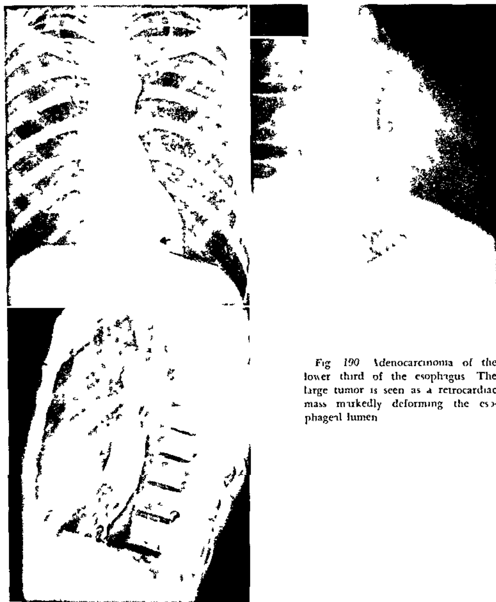


Fig 190 Adenocarcinoma of the lower third of the esophagus The large tumor is seen as a retrocardiac mass markedly deforming the esophageal lumen

fication as mediastinal masses but smaller ones may not be so apparent Esophageal tumors encroaching on the trachea may also cause visible deformities of that structure

The benign tumors in general show larger mediastinal deformities than those *of a malignant nature* They are usually rounded or oval and smooth or lobulated The majority being leiomyomas are found in the middle and lower thirds of the esophagus where this tumor most commonly grows

Schatzki and Hawes have described the radiologic characteristics of these benign tumors They state When studied with a contrast medium the mucosa of the esophagus is stretched over the tumor and the folds are obliterated In profile view there is an abrupt angle between the normal and *the involved esophagus this angle being* sharpest when the mass is nearest the lumen but can also be sharp when the mass is at some distance from the lumen but attached to the esophagus In face on view the edge of the lesion lies transversely or obliquely and is sharply outlined The esophageal luminal defect is smooth or lobulated but not irregular The size of the esophageal defect is indicative *of the tumor size The esophageal defect* may be inconstant when the lesion is small being affected by the esophageal filling and the plane of visualization The esophageal attachment of the lesion can be confirmed by the fact that it moves with swallowing

Schatzki and Hawes have also discussed the differential diagnosis of intrinsic and extrinsic mass lesions in and around the esophagus and conclude the following *(1) It is usually easy to differentiate* lesions which are extrinsic to the esophagus and not attached from those which either arise in the wall of the esophagus or have become secondarily attached to this structure (2) It is difficult to differentiate between mucosal lesions extramucosal lesions and extrinsic *lesions attached to* the esophagus

The malignant tumors of the esophagus are generally smaller in *size than the benign* lesions Haas and Baker point out that their soft tissue outlines are best seen in lateral and right anterior oblique projections but for more complete study other projections are indicated as well They state that progression and regression of an individual lesion can be studied by this method

Examinations with contrast media however *reveal the extent of the luminal deformity* The characteristics of the deformity are determined by the pattern of growth In the fungating type there are sharp shelving margins between the normal and involved esophagus and irregularities of the borders of the narrowed lumen (because of mucosal destruction) In the scirrhous type the tumor constricts the esophageal lumen in an hour glass *fashion and the mucosa is usually smooth* In the polypoid type there are multiple lobular defects in the lumen commonly extending over a relatively long segment of the esophagus

Benign and malignant tumors of the esophagus rarely contain visible calcium

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Chapter 24

DIAPHRAGMATIC HERNIAS

DIAPHRAGMATIC hernia according to Harrington is a term commonly used to designate any condition in which there is protrusion of abdominal contents into the thoracic cavity through an abnormal opening in the diaphragm There are many *classifications of these herniations* based on such categories as pathologic anatomy etiology the diaphragm opening involved the content of the hernia whether congenital or acquired whether traumatic or nontraumatic and whether or not covered by herniated peritoneum in the latter instance those which lack a peritoneal covering might more appropriately be called eviscerations false hernias or pseudohernias

In this text only those herniations commonly projecting into the mediastinum will be considered these are (1) esophageal hiatus hernias and (2) foramen of Morgagni hernias Other types may occasionally involve the mediastinum notably large foramen of Bochdalek herniations midline diaphragmatic ruptures by trauma and inflammation and herniations through congenital midline defects in the diaphragm

ESOPHAGEAL HIATUS HERNIAS

Esophageal hiatus hernias are abnormal protrusions of abdominal contents into the chest through the esophageal hiatus Kirklin and Hodgson estimate that 98 per cent of all diaphragmatic hernias are of this nature The stomach is the most common organ involved in such herniations but occasionally the colon small intestine omentum and other structures may herniate in a similar way At times several structures may be involved

The gastric herniations have been divided into several types depending upon the length of the esophagus and the relationship of the lower esophagus to the herniated portion of the stomach All of these result in mass formations in the lower mediastinum which are more or less *indistinguishable on ordinary chest examinations* but which can be differentiated by contrast media studies

The herniated mass is composed of several structures these include the herniated sac of peritoneum the gastric wall and the contents in the lumen of the herniated stomach (usually a mixture of air and fluid) Grossly and microscopically the wall of the stomach may either be perfectly normal or may have a varying degree of inflammation ulceration and congestion The herniated sac is usually located in or near the mid line of the body this is in

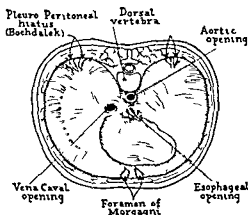


Fig 191 Sketch of diaphragm seen from above showing position of openings between the chest and abdomen From Caffey J *Pediatric X ray Diagnosis* Third Edition page 22 Chicago Ill The Year Book Publishers Inc 1956

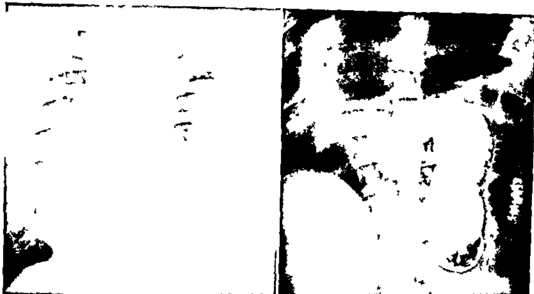


Fig 192 Traumatic rupture of the diaphragm with herniation of the stomach entire small intestine and part of the colon into the mediastinum and left pleural space The diaphragmatic defect measured 12 cm in diameter

contrast to the epiphrenic diverticulum of the esophagus which is laterally placed usually on the right

Radiologically, esophageal hiatus hernias are frequently first detected on routine chest examinations where they are seen as midline retrocardiac masses The most important point in their identification and differentiation from other lower mediastinal masses is the presence of air fluid levels on films which have been made with the patient in upright (or decubitus) positions For the identification of such levels over penetrated films are advisable thus



Fig 193 Esophageal hiatus hernia The large midline gastric mass can be seen through the heart shadow in the frontal view Air and fluid (arrows) are present within the lumen



Fig 191 Esophageal hiatus hernia An air fluid level in the herniated stomach can be seen in the right mediastinum on the frontal view (arrow) but is better visualized on the lateral projection

holds particularly true for the frontal view where the mass is obscurred to some extent by the superimposed heart and thoracic spine Absence of air fluid levels in upright (and decubitus) views does not rule out the diagnosis of herniation since in many instances the sac may be collapsed or may be completely filled with fluid

Examination of the gastrointestinal tract with contrast material proves the diagnosis In such studies several points can be determined these include (1) the size of the mass (2) the structures contained in the mass (stomach duodenum small intestine colon etc) (3) the relationship of the lower esophagus to the hernia (4) the presence or absence of ulcerations tumors or other lesions in the lower esophagus or herniated stomach (5) the presence or absence of gastroesophageal reflux (6) the diameter of the diaphragmatic opening (7) the presence or absence of obstruction at the esophagogastric juncture and at the esophageal hiatus and (8) the appearance of the intraabdominal portion of the stomach and the duodenum

When herniations of small intestine or colon occur through the esophageal hiatus the correct diagnosis may be suspected on routine roentgenologic examinations of the chest by the appearance of the intestinal patterns

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Fig 192 Esophageal hiatus hernia with the entire stomach (air containing mass above diaphragm to left) duodenum and splenic flexure area of the colon (opacified here with barium) in the hernial sac in the mediastinum

and related problems (Editorial) *Radiology* 62 750 1954
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FORAMEN OF MORGAGNI HERNIAS

The foramina of Morgagni are small triangular shaped congenital defects in the left and right sides of the diaphragm anteriorly and medially adjacent to the sternum Lymphatics and the superior epigastric vessels course through them Superiorly they are covered by pericardium inferiorly by peritoneum

Occasionally they are enlarged allowing *abdominal contents to herniate into the chest* this occurs more commonly on the right side than on the left where the pericardium and heart tend to protect the opening In rare instances the two foramina are joined creating one large foramen this is due to a congenital absence of the usual midline attachment of the *central tendon of the diaphragm to the* posterior aspect of the sternum

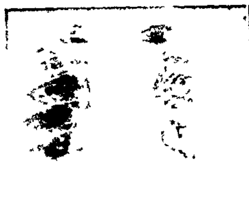
Several names have been given to herniations of this type including substernal parasternal costosternal subcostosternal hernia and hernia of Larrey s space The term foramen of Morgagni hernias is the one preferred by the authors

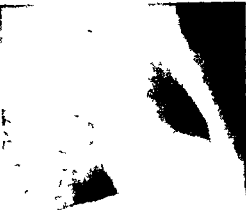
These herniations are thought to be both congenital and acquired They are usually true hernias in that the herniated structures are contained within a peritoneal sac The most frequent structures involved are the omentum colon stomach and small intestine In one reported series of 27 such cases the colon alone was herniated in 8 the omentum alone in 4 and the colon and omentum in 5 in the remainder there were such structures as stomach small intestine liver and spleen involved Omental herniations are more frequent in the older age group (especially in women who have borne children) whereas alimentary tract herniations are more common in the younger individual

Grossly the herniated masses are rounded or oval in shape and smooth They vary



Fig 196 Foramen of Morgagni hernia in a 9 year old school boy Asymptomatic This left sided hernia contained omentum liver and bowel in a peritoneal sac





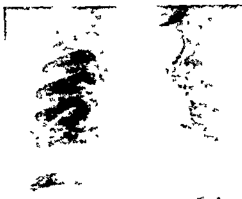






Fig. 197. Right foramen of Morgagni hernia of omentum. The chest examination in frontal (A) and lateral (B) projections discloses the rounded orange-sized mass in the right cardiophrenic angle. Following pneumoperitoneum, upright frontal (C) and lateral (D) chest films show air filling the upper half of the herniated peritoneal sac (with omentum in the lower half) and air under each diaphragm leaf. Barium enema examination (E) reveals a displacement of the transverse part of the colon toward the hernia.

in size but the usual ones are about that of an orange or lemon The same mass may vary at times due to some degree of sliding between the chest and abdomen (usually the result of alterations in pressure between the two areas) The hernias are located anteriorly and medially and are bounded by pericardium lung anterior chest wall and diaphragm On the abdominal side the round and falciform ligaments of the liver are usually in intimate relationship with the neck of a sac

Radiologically, on routine frontal and lateral chest examinations these herniations are seen as rounded or oval shaped masses resting on the diaphragm in the cardiophrenic angles The ones on the left may displace the heart to the right Those on either side may compress adjacent lung tissue When the mass is of homogeneous density it may contain such structures as omentum liver spleen or non air containing bowel or stomach Air in a sac indicates that some type of hollow viscus is present (providing there is no pneumoperitoneum)

Fluoroscopic observations in various positions and stages of respiration may show that the hernia changes in size and shape during such maneuvers and that it cannot be separated from the diaphragm Transmitted pulsations from the heart may be apparent

In the radiologic study of these and other types of cardiophrenic angle masses examinations of the gastrointestinal tract are exceedingly important Such examinations may show that a part of the colon small intestine or stomach are either within the mass or have been pulled toward the hernia by omentum which is incorporated in the sac The absence of such displacements however is of no diagnostic significance

Foramen of Morgagni hernias can generally be diagnosed following pneumoperitoneum Films of the chest made in upright position following such a procedure will show the presence of air in the herniated peritoneal sac Absence of air from the sac following pneumoperitoneum generally means either faulty technique or obstruction of the sac s neck at the diaphragm In the occasional instance where the herniated structures lack a peritoneal covering (false sac) a pneumothorax or pneumomediastinum may result when a pneumoperitoneum is done

Omentum being composed of fat casts a shadow of lesser density than other soft tissues but this is difficult to evaluate in the cardiophrenic angles because of the superimposed air in the lungs The other fat containing masses of the mediastinum are the epipericardial fat pads teratomas of the dermoid type lipomas and liposarcomas

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Chapter 25

LESIONS OF BONE AND CARTILAGE

THERE ARE several different types of bone and cartilage lesions which affect the mediastinum These include the tumors of bone and cartilage vertebral inflammations with associated paravertebral abscesses scoliosis and osteophytosis Each of these will be discussed separately in this chapter

In addition to abscesses of vertebral origin the mediastinum is also affected by abscesses from other causes such as trauma perforations and spread of inflammation These types of abscesses do not belong in this section and so are discussed in the chapter entitled Mediastinitis

TUMORS OF BONE AND CARTILAGE

Bone and cartilage tumors located at thoracic levels not uncommonly produce mediastinal tumefactions The majority of these are benign or malignant tumors arising from the dorsal vertebrae but occasionally tumors arise from the ribs adjacent to the spine from the clavicles sternum costal cartilages the tracheal cartilages and possibly even from cartilage implants in the mediastinum

Tumors of the dorsal spine may produce mediastinal masses in several different ways as follows

(1) The tumor may grow more or less eccentrically off the vertebral body or pedicle *resulting in a localized mass*

(2) The tumor may grow within the body of the vertebra expanding it but remaining confined within its cortical boundaries

(3) The tumor may arise in the vertebra initially expanding it but eventually breaking through into the soft tissues resulting in a paravertebral tumefaction

(4) The tumor may arise in the vertebra destroy the body and pedicle partially or completely and extend into the paravertebral area as a soft tissue mass

At times these paravertebral soft tissue masses mentioned above may be composed of pus or hemorrhage as well as of tumor tissue

In some instances a tumor may appear in the paravertebral soft tissues without any obvious bone pathology these may be vertebral or rib tumors with undetected destruction periosteal (or other fibrous tissue) tumors metastatic implants in the paravertebral soft tissues or spinal canal tumors extending into the paravertebral soft tissues

The most commonly encountered benign neoplasms protruding into the mediastinum from the thoracic spine sternum ribs or clavicles are osteochondromas and aneurysmal bone cysts Less frequently seen are giant cell tumors Though hemangiomas are commonly observed in vertebrae significant expansion of this tumor into the mediastinum is only rarely noted

The more important primary malignant tumors of bone or cartilage extending into the mediastinum are chondrosarcomas osteogenic sarcomas and fibrosarcomas Other noteworthy malignant tumors arising in bone and simulating mediastinal masses are the lymphomas Ewing's sarcomas *reticulum cell sarcomas and myelomas Rarely* chordomas originate from the thoracic spine and bulge into the mediastinum

Undoubtedly bone metastases are the most common osseous lesions producing mediastinal tumefactions Mass formations secondary to metastases may pose considerable difficulties in differential diagnosis from many types of mediastinal lesions

Radiologically, since most bone and cartilage tumors of the mediastinum arise

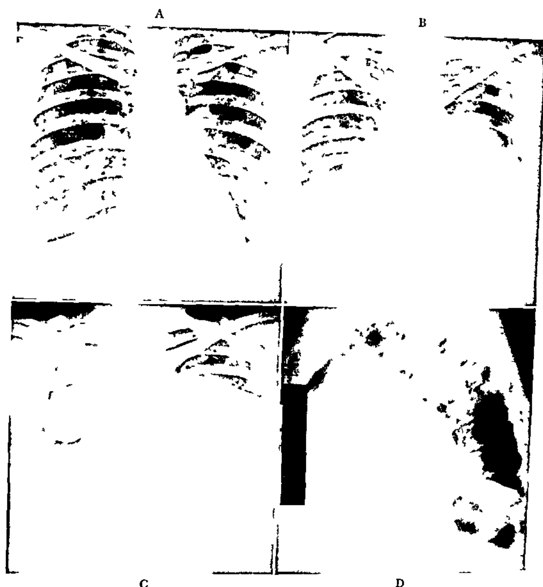


Fig 198 Chondromyxosarcoma of the anterior mediastinum in a 48 year old female Frontal views of the chest (A B & C) show extremely rapid growth of the tumor over a period of about 2 months The lateral view (D) shows the anterior position of the mass Post mortem examination revealed that the tumor had extensively invaded the main pulmonary artery and some of its branches in the lungs and was adherent to and invading the pericardium The tumor's origin was indefinite but development from a basic undifferentiated cell was postulated From Schwinger A and Hemley S D Anterior mediastinal chondromyxosarcoma *Dis Chest 21* 670 1953

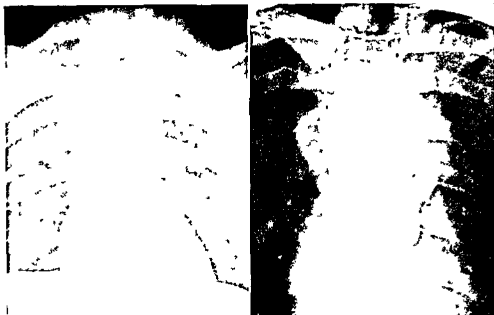


Fig. 199 Chondromyxosarcoma of the posterior mediastinum arising from a dorsal vertebra. A frontal chest film discloses a small mediastinal mass. A bucky film of the same area reveals destruction of the vertebral body and a soft tissue tumefaction. Surgical exploration disclosed the malignant tumor of the vertebra and its extension into the paravertebral space. (Courtesy of Simon Krantz, M.D., V. A. Hospital, Atlanta, Ga.)



Fig. 200 Osteochondroma of the right sixth rib arising retropleurally. Dense bony deposits occupy the larger portion of the tumor.



Fig 201 *(See Next Page)*

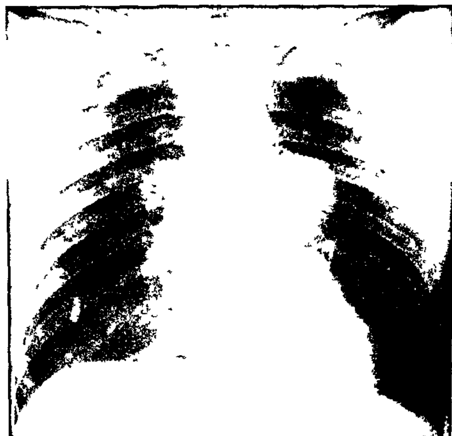


Fig 201 Chondrosarcoma of the mediastinum with involvement of the spinal canal (hour glass tumor) *Frontal and oblique roentgenograms of the chest reveal a sharply circumscribed* mass in the posterior mediastinum with involvement of the adjacent ribs On surgical exploration the tumor was also found to involve the adjacent vertebrae and epidural space From Weisel W and Ross W B Chondrosarcoma of the posterior mediastinum with hourglass involvement of the spinal canal resection and recovery *J Thoracic Surg* 19 645 19[illegible]0



Fig 202 Chordoma arising from a dorsal vertebra causing a distinct soft tissue mass From Wood E H Jr and Himadi G M Chordomas A roentgenologic study of sixteen cases previously unreported *Radiology 54* 706 1950



Fig 203 Multiple myeloma with involvement of the tenth dorsal vertebra A mediastinal mass has resulted from bilateral extension of the tumor into the paravertebral soft tissues

from the thoracic spine or the adjacent ribs it is logical that most of them are confined to the posterior mediastinum or to the posterior part of the superior mediastinum Occasionally tumors (notably chondromas) arise from other bone and cartilage bearing structures in or bordering the mediastinum (sternum clavicles rib cartilages trachea) and these may be located further forward in the mediastinum At other times a skeletal tumor may become so large that its exact origin becomes indefinite both radiologically and surgically For practical purposes the discussion here will be based principally on those lesions which arise from the thoracic spine and from those portions of the ribs adjacent to the spine

Adequate radiologic investigation is extremely important in bone and cartilage tumors The following points should be determined in any one examination (1) The location of the lesion within the mediastinum its size and shape (2) the number of vertebrae (or ribs)) involved (3) the characteristics of the vertebral (or costal) destruction and/or proliferation (4) the presence or absence of calcium in the lesion and the characteristics of the deposits



Fig 204 Ewing s endothelioma of the seventh dorsal vertebra in a 10 year old boy The left side of the vertebral body and pedicle are partially destroyed and there is extension of the tumor into the paravertebral soft tissues From Epstein B S *The Spine* Philadelphia Lea & Febiger 1955

when present (5) the presence or absence of an associated paravertebral mass and the characteristics of this mass (6) evidences of spinal cord or peripheral nerve involvement (7) evidences of similar lesions elsewhere in the spine or other bones of the body and (8) evidences of metastatic spread to the soft tissues Taking all these points into consideration and knowing the general characteristics of the different neoplasms that can involve the dorsal spine and ribs one can often narrow the diagnostic possibilities down to several entities unfortunately a specific diagnosis is rarely possible when based on radiologic characteristics alone

Paravertebral soft tissue masses when associated with bone and cartilage tumors may be composed of tumor tissue necrosis or hemorrhage or to combinations of these Generally these soft tissue extensions are either unilateral or asymmetrically bilateral—a point of value in differentiating them from soft tissue masses on the basis

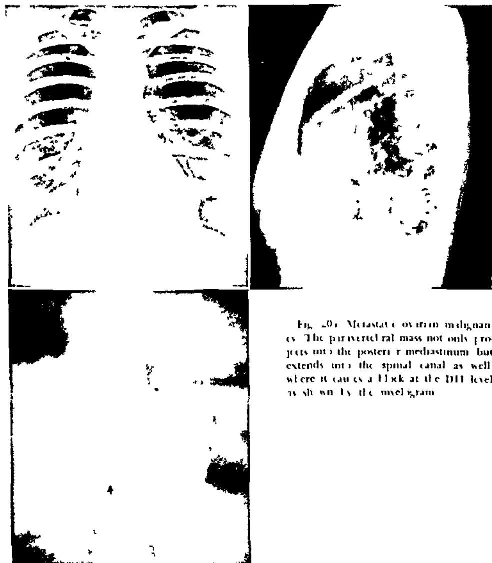


Fig. 201 Metastatic ovarian malignancy The paravertebral mass not only projects into the posterior mediastinum but extends into the spinal canal as well where it causes a block at the D11 level as shown by the myelogram

of osteomyelitis of the spine where the lesions are often bilateral and symmetrical. Paravertebral masses may be confined to the level of the tumor in the spine or may extend for a variable distance above and below it; in most all cases, however, the broadest part of the soft tissue mass corresponds to the area of greatest destruction in the spine.

The visible borders of these paravertebral masses are seen best in frontal projection although many times they are visible also in the obliques and lateral views. Frequently in the frontal projection similar lines of other mediastinal structures cause difficulties in identification; these notably are the descending aorta on the left and the ascending aorta and superior vena cava on the right. Tomography in the frontal projection is often helpful in these cases. (The normal paravertebral lines are discussed in the section on anatomy.)

The importance of a skeletal survey cannot be stressed too much for a surprising number of clinically silent lesions will be uncovered in this way and their presence will aid in a differential diagnosis.

Myelography serves the purpose of identifying the levels and degrees of spinal cord compression and spinal canal blockage and in identifying any other levels which may be involved.

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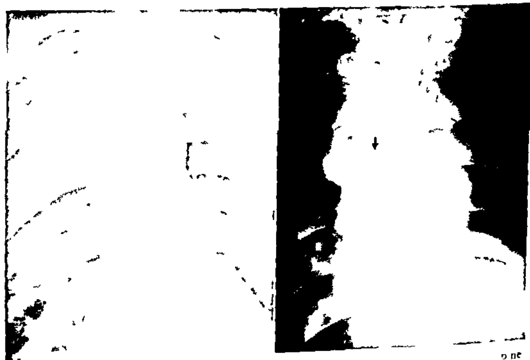
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VERTEBRAL INFLAMMATIONS AND ASSOCIATED PARAVERTEBRAL ABSCESSES

Paravertebral abscesses are in most instances the result of spinal osteomyelitis occasionally they are due to neoplasms of the vertebrae and to extensions from abscesses in the spinal canal In this discussion consideration will be given not only to the abscess but to the vertebral osteomyelitis as well

In most instances the bacteria causing osteomyelitis are blood borne to the spine An explanation for this is forthcoming through the experiments of Batson who showed that the vertebral venous system which consists of a network of valveless blood vessels richly communicates with the other veins of the body cavities at various intervertebral levels and that foreign elements within the blood stream such as bacteria and neoplastic cells may reach the vertebrae quite easily

Spinal osteomyelitis usually begins in the spongiosa of the vertebral body near the upper or lower vertebral plates anteriorly



Fig 207 Paravertebral abscess secondary to tuberculosis of the thoracic spine The lesion involves 2 vertebrae and the disc between them The abscess extends predominantly to the right



Fig 208 Non tuberculous osteomyelitis with marked vertebral destruction demonstrated by a tomographic study of the thoracic spine

The spongy bone is rather quickly replaced by an abscess pocket and as the destructive process spreads the adjacent intervertebral disc and vertebra are involved Spread of the inflammatory process into the soft tissues at this time results in a paravertebral abscess this abscess is located anteriorly and on the sides and is greatest in extent at the level of the lesion

Further spread of the inflammation results in additional manifestations Other adjacent vertebrae and discs may become involved or similar lesions may arise at other levels in the same segment or other segments Angulations of the spine occur as a result of the destruction and wedging of the vertebral bodies The paravertebral mass becomes larger and because of gravitation of the pus may become broader in its lower portions Calcifications may appear in the mass

On healing the paravertebral mass may disappear partially or completely Two or more adjacent vertebrae which have been partially destroyed may fuse into a single bony mass Partially destroyed vertebra may become sclerotic in the remaining por

tions Permanent angulations of the spine may result

Most cases of osteomyelitis are either tuberculous or pyogenic Generally the spinal involvement in the two types is more or less the same but differences do exist in certain aspects and these will be mentioned

Spinal tuberculous osteomyelitis according to Schinz *et al* is the most frequent type of skeletal tuberculosis in adults the second most frequent in children The course of the inflammation as compared with the pyogenic types is usually longer Vertebral destruction is often marked Sharp angulations of the spine (humps) are relatively frequent and often severe Bony sclerosis is occasionally seen Paravertebral abscesses are common and are often very large when they become large, they are likely to be gravitational in type Multiple levels of involvement are infrequent

Pyogenic spinal osteomyelitis is more common than realized Recently several authors (Henson and Coventry Sherman and Scheider Leigh *et al*) have pointed up its frequent association with infections in the urinary tract and in other pelvic organs Vertebral destruction in pyogenic osteomyelitis is relatively less marked than with tuberculosis and residual spinal deformities are minimal Bony sclerosis occurs commonly Paravertebral masses are common but are usually smaller tend to remain localized to the site of the lesion and are not likely to be gravitational in type Multiple levels of involvement are frequent

Radiologically, when a paravertebral mass is disclosed the adjacent spine as well as the mass should be adequately studied Often one can arrive at a differential diagnosis between inflammation and neoplasm

In a study of paravertebral abscesses several points should be determined These are as follows (1) The extent of the abscess in the paravertebral soft tissues with accurate estimations of its upper most and lowest limits (2) the relation of the soft tissue mass to the vertebral lesion whether greatest in width at the lesion or below it (3) the presence or absence of calcium deposits within the soft tissues and (4) evidences of encroachment on the spinal cord or peripheral nerves using myelography if indicated

In studying the lesion within the spine it is important to know the following points (1) The extent of involvement of the vertebra (2) the number of vertebrae and discs involved (3) the characteristics of the destroyed area (4) the presence or absence of other levels of involvement in the spine and (5) evidence of involvement of other bones of the body or of lesions in the lungs

The value of tomography in the study of vertebral lesions cannot be overemphasized This special procedure is particularly helpful in identifying small abscesses and in differentiating these from slight widenings of the paravertebral lines caused by osteophytes in differentiating abnormal paravertebral lines from other normal contour lines of the mediastinum and in studying the inflammatory lesions in the spine itself particularly small areas of destruction which may not be visible by other means

Serial examinations of the lesions in the spine and paraspinal areas affords an opportunity to determine when the lesions seem to be further progressing static or regressing

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SCOLIOSIS

Scoliosis of the thoracic spine is exceedingly common it may result from many causes both congenital and acquired The predominant curve in scoliosis is to the right and involves a variable segment of the spine There may be associated curves in the cervical and lumbar segments either to the same side or contralaterally

Radiologically, scoliosis may simulate a soft tissue mediastinal tumefaction in the frontal view if the film density is not sufficient to show the bony details of the spine This diagnosis should always be considered in lightly exposed films when there is a smooth broadening of the mediastinum to the right or left With thoracic scoliosis there is invariably asymmetry of the left and right rib cages and this finding if present should immediately bring to mind the correct diagnosis

Adequately exposed films offer no problem in diagnosis the spine can be seen and identified In the occasional case there may be a mediastinal mass present in a patient who also has a scoliosis of the thoracic spine such a lesion may be missed unless adequate radiologic study in multiple projections is made (See chapter on neurogenic lesions)

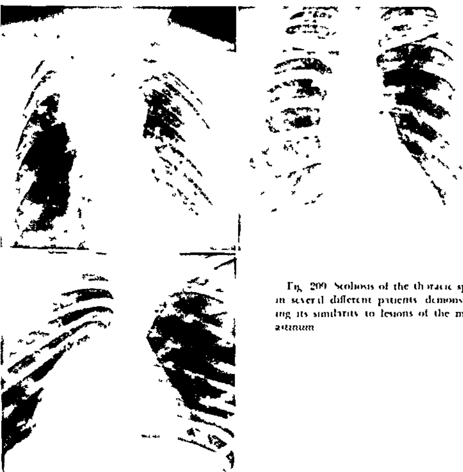


Fig. 200 Scoliosis of the thoracic spine in several different patients demonstrating its similarity to lesions of the mediastinum



Fig 210 Localized osteophytosis of the lower thoracic spine resembling a small mediastinal mass in the frontal projection

OSTEOPHYTOSIS

Osteophytosis is perhaps the most common lesion of the thoracic spine and is a manifestation of disc damage The spurs are most frequent on the right and anterior sides of the vertebral bodies at their margins adjacent to the disc spaces they are less common on the left side of the bodies (probably because of the pulsation of the descending thoracic aorta) and are rare posteriorly A variable number of vertebrae may show osteophytosis

Radiologically these bony spurs may resemble mediastinal tumefactions in the frontal view particularly when there is insufficient exposure of the film to show bone detail and when the spurs are localized to one level Adequate exposure of the films should establish the diagnosis Osteophytes on the anterior sides of the vertebrae are best seen on lateral film studies and should offer no difficulty in identification in films made with the usual techniques

Chapter 26

LESIONS OF THE AORTA AND ITS BRANCHES

IN THIS CHAPTER the principal congenital and acquired lesions of the aorta and its branches are described. Pathologic changes of the aorta and its large vessels frequently alter the radiologic appearance of the mediastinum or cause significant deformities of the mediastinal viscera. In many instances the radiologic abnormalities produced by vascular lesions require their consideration in the differential diagnosis of various types of mediastinal diseases and mass formations.

CONGENITAL ANOMALIES

Vascular malformations of the superior mediastinum are of clinical importance on account of their compression effects on the trachea and esophagus or arterial blood flow changes. The radiologic examination of the mediastinum with and without the use of contrast media is a most important means of recognizing these conditions.

There are many possible derangements of the aortic arch system and large vessels. Only the most frequent anomalies shall be described here. A proper understanding of the anatomic abnormalities occurring in these vascular derangements makes it possible to recognize also those malformations which are not presented in this chapter.

Double Aortic Arch. In this abnormality both the right and left aortic arches are persistent. The left arch which is located anterior to the trachea is usually the smaller one and both arches surround the trachea and esophagus in a ring-like manner. The arches fuse behind the trachea and esophagus to form the descending aorta which is usually located on the left side but which in some instances will be found in front of or toward the right of the dorsal spine. Though the double aortic arch may not cause clinical symptoms, difficulties of respiration and swallowing are frequently observed in this abnormality. Infants with this abnormality may have a labored respiration; there may be marked inspiratory and expiratory stridor accentuated during swallowing. Respiratory infections in these infants and young children are common.

On radiologic examination in the frontal projection the ring-like double aortic arch produces an indentation of the lateral walls of the distal third of the trachea. Though this may be recognized on overexposed film studies, contrast examination of the trachea will usually demonstrate this bilateral defect to advantage. In the lateral view the distal third of the trachea shows an anterior indentation resulting in marked narrowing of the lumen. The esophagus at approximately the same level shows along its posterior aspect a large extraluminal defect corresponding to the diameter of the right posterior arch. This defect is rather horizontal and may occasionally show pulsation. Angiocardiography is rarely necessary to demonstrate this malformation.

Right Aortic Arch. In this anomaly only the right aortic arch system persists with obliteration of all or part of the left aortic arch. The aortic arch is found along the right lateral aspect of the trachea and the esophagus. The descending aorta may be found either to the right or to the left of the dorsal spine. Usually a right-sided aortic arch will not cause any untoward compression symptoms of the esophagus and trachea.

On radiologic examination the entire trachea is found to be displaced toward the left side and the main soft tissue shadow of the mediastinum is found to the rig

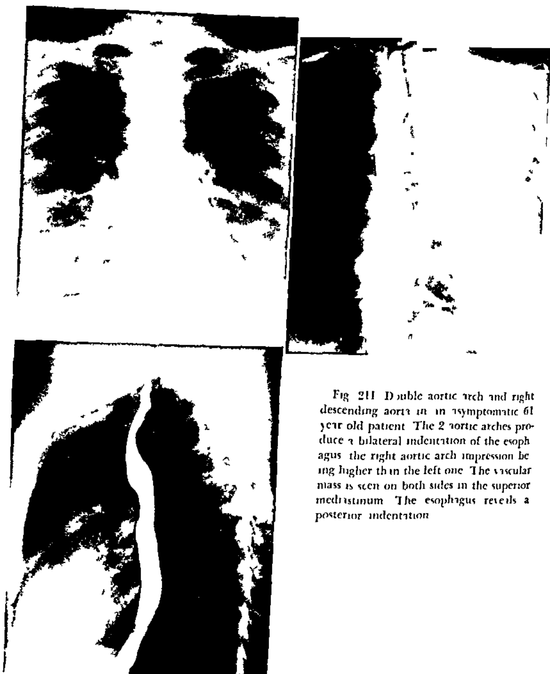


Fig 211 Double aortic arch and right descending aorta in an asymptomatic 61 year old patient The 2 aortic arches produce a bilateral indentation of the esophagus the right aortic arch impression being higher than the left one The vascular mass is seen on both sides in the superior mediastinum The esophagus reveals a posterior indentation

pect of the trachea towards the right side of the neck. In a similar way a left carotid artery arising somewhat more proximal than usual will have to pass along the anterior aspect of the trachea towards the left side of the neck. In both abnormalities respiratory stridor may occur due to anterior tracheal compression.

On radiologic examination the anterior tracheal compression may be recognized on lateral chest roentgenogram but should be confirmed by examination of the trachea with contrast media. In this abnormality no esophageal indentation or displacement will be recognized. These abnormalities have to be differentiated from a primary softening or underdevelopment of the tracheal cartilages by observation of the trachea during the entire respiratory cycle. In the abnormalities described above the defect is usually persistent whereas a softening of the trachea will show variation in the tracheal lumen during respiration.

Aberrant Right Subclavian Artery. The right subclavian artery may arise directly from the aortic arch distal to the origin of the left subclavian artery. Under these circumstances the right subclavian artery will pass to the right side behind the esophagus. In very rare instances pressure of the artery on the posterior aspect of the esophagus may lead to dysphagia.

The aberrant subclavian artery will not produce any changes in the contour of the mediastinum on conventional radiologic examination. On contrast examination of the esophagus an oblique shelf-like filling defect at or above the level of the aortic arch may be detected. The small diameter of this defect differentiates it from the

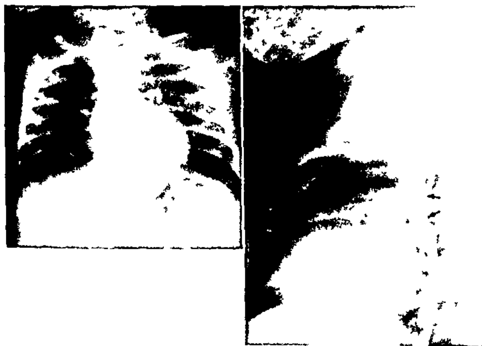


Fig. [illegible]11. Vascular ring due to right aortic arch and left ligamentum arteriosum. In the frontal roentgenogram of the chest the main soft tissue mass of the superior mediastinum is seen on the right side indicating the presence of a right-sided aortic arch in this patient with respiratory distress. The lateral tracheogram and esophagogram reveal a sharp angular posterior indentation by the ligamentum arteriosum which extends from the left pulmonary artery to the descending aorta on the right side (arrows).

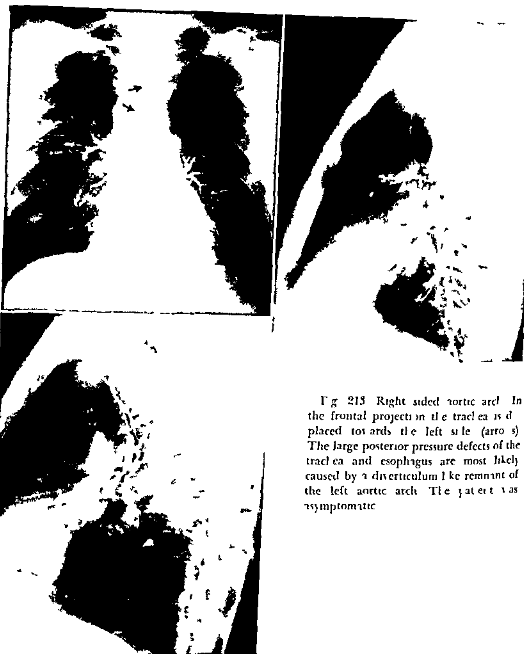


Fig 213 Right sided aortic arch In the frontal projection the trachea is displaced towards the left side (arrows) The large posterior pressure defects of the trachea and esophagus are most likely caused by a diverticulum like remnant of the left aortic arch The patient was asymptomatic

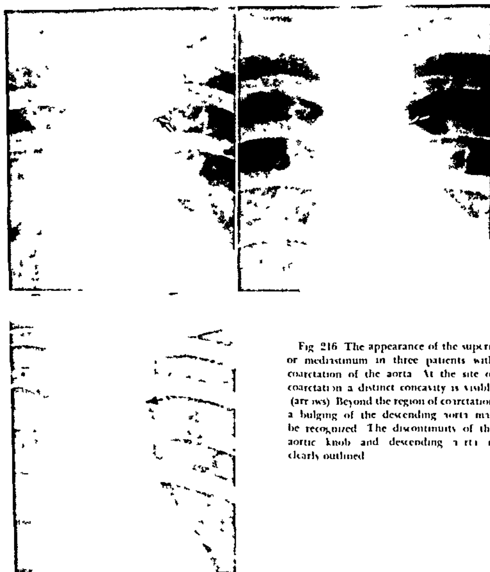


Fig 216 The appearance of the superior mediastinum in three patients with coarctation of the aorta At the site of coarctation a distinct concavity is visible (arrows) Beyond the region of coarctation a bulging of the descending aorta may be recognized The discontinuity of the aortic knob and descending aorta is clearly outlined

presence of a double aortic arch. Frequently this defect is better demonstrated if only a small amount of contrast medium is administered. The defect is so characteristic that it can hardly be mistaken for a neoplasm.

Coarctation of the Aorta. Coarctation is a faulty development of the aorta in the vicinity of the entrance of the ductus arteriosus. In most cases of coarctation narrowing is found a short distance beyond the origin of the left subclavian artery. In the adult type of coarctation the ductus arteriosus enters the aorta proximal to its constriction, whereas in the infantile type *the ductus arteriosus passes to the aorta* distal to the narrowing. Lack of collateral circulation in the latter type of coarctation makes survival into later periods of age uncommon. *A significant anatomic feature* of this condition is a forward and medial buckling of the aorta at the site of constriction and insertion of the ligamentum arteriosum. Below the site of constriction a dilatation of the aorta is frequently observed which in some instances has assumed aneurysmal proportions. Many cases of coarctation are associated with aortic cusp anomalies. Death from this malformation may be due to left ventricular failure as the result of hypertension or rupture of the aorta or dissecting aneurysm.

The radiologic features of coarctation of the aorta are often characteristic and may be demonstrated with conventional x-ray procedures.

In many cases of coarctation a shallow left concave indentation of the aorta appears below the contour of the knob. Distal to this indentation a slight outward bulge of the descending aorta may be recognized, often larger than expected in the *age group of the patient. Normally the* shadow of the aortic knob blends gradually without interruption with that of the descending aorta. In coarctation a certain *discontinuity of the aortic knob and the* descending aorta is often noted. The aortic knob appears frequently abnormal in contour, size and position. Surgical and angiocardiographic observations have disclosed that the apparent knob is in some instances formed by a bulbous dilatation of the most proximal segment of the dilated left



Fig. 215. Aberrant right subclavian artery. The aberrant right subclavian artery arises as the last large vessel from the aortic arch and courses behind the esophagus to the right side. The esophagus shows an oblique posterior pressure defect (arrows).

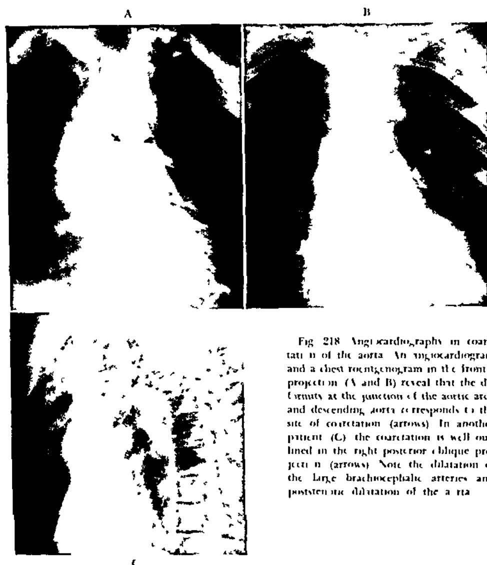


Fig 218 Angiocardiography in coarctation of the aorta An angiocardiogram and a chest roentgenogram in the frontal projection (A and B) reveal that the deformity at the junction of the aortic arch and descending aorta corresponds to the site of coarctation (arrows) In another patient (C) the coarctation is well outlined in the right posterior oblique projection (arrows) Note the dilatation of the large brachiocephalic arteries and poststenotic dilatation of the aorta

subclavian artery There are probably several factors which contribute to the radiologic appearance of the abnormal aortic knob in coarctation of the aorta such as the buckling of the aorta the dilatation and elongation of the proximal aorta and subclavian arteries

In the left anterior oblique position the shadow of the descending aortic arch and aorta may be invisible *This finding is not* necessarily diagnostic as the descending aorta is frequently not recognized in normal children In about one third of adult patients an indentation of the posterior superior contour of the aorta may be recognized corresponding to the site of coarctation Tomographic studies may demonstrate this finding to advantage On fluoroscopy the ascending aorta is often seen to be prominent with vigorous pulsations whereas pulsations in the descending aorta are absent Above the level of the aortic knob the dilated actively pulsating large brachiocephalic arteries are commonly recognized

Normally the aorta below the summit of the arch is found in close relationship to the left lateral aspect of the esophagus As coarctation occurs in this area deformity of the esophagus may be expected In coarctation the esophagus below the level of *the knob is frequently displaced to the* right and anteriorly probably due to the kinking of the aorta and dilatation beyond the site of constriction The impression by the aortic arch is often found to be very *small*

The tortuous intercostal arteries in coarctation produce a more or less distinct notching of the inferior borders of the ribs in approximately three fourths of all affected individuals A well developed rib notching is almost always due to coarctation of the aorta Among the rarer causes



Fig 217 *Displacement of the esophagus in coarctation of the aorta* In the frontal *projection* a slight right lateral displacement of the esophagus at the level of the *postconstriction* dilatation of the descending aorta is visible This aortic dilatation is also demonstrated by anterior displacement of the esophagus in the left anterior oblique view (arrows)

of various diseases affecting this blood vessel

The principle causes of pathologic changes in the wall of the aorta are the following

(1) Senile ectasia
(2) Atherosclerosis
(3) Aortitis
(4) Hypertension
(5 Aortic valvular lesions

In senile ectasia the elastic elements of the aorta have been overstretched and damaged resulting in permanent increase of the length and width of this blood vessel The loss of elasticity is probably the result of physical alterations of the elastic fibers with the ageing process and is frequently associated with extensive connective tissue proliferation in the aortic wall It has been postulated that the fibrosis may to some extent compensate for loss of elasticity and provide increased strength and rigidity of the vessel Senile ectasia usually occurs only in mild forms and pronounced dilatation and elongation of the aorta on this basis alone is rather infrequent

Atherosclerosis is predominantly a disease of older age groups though it is not unusual to find it in the young The early pathologic changes are characterized by development of fibrosis and lipoidosis in the internal layers of the aorta The fatty deposits accumulate in circumscribed plaques which may be slightly elevated and protrude into the lumen of the blood vessel Subsequently the atheromatous patches may calcify and ulcerate leading not infrequently to the formation of thrombi Atherosclerosis affects predominantly the immediate supravalvular area of the aorta close to the coronary ostia the aortic arch in the neighborhood of the orifices of the brachiocephalic vessels and the descending aorta It is noteworthy that the ascending aorta shows comparatively little involvement in contrast to luetic aortitis which favors this segment

Infectious diseases of the aorta are predominantly syphilitic in character though other inflammatory processes such as rheu



Fig 219 Elongation of the aorta in hypertensive heart disease The aortic knob is elevated and appears at the level of the clavicles The distance from the right cardiovascular junction to the aortic knob is increased There is also tortuosity of the innominate artery present (arrows)

matic infections are commonly observed on pathologic examination The syphilitic infection of the aorta produces a characteristic periaortitis and mesaortitis with secondary fibroblastic changes of the intima Extensive necrosis of the media may take place with marked destruction of its musculoelastic elements This process is predominantly found in the ascending aorta beginning just above the level of the aortic cusps

The elongation of the aorta in hypertension is frequently a purely dynamic one Inasmuch as hypertension is commonly associated with atherosclerosis structural changes of a more permanent character may take place in the course of hypertensive heart disease

Some degree of elongation and dilatation of the aorta is commonly observed in patients with aortic valvular lesions If aortic insufficiency has developed on a luetic basis,

of rib notching are pulmonary stenosis or atresia intercostal vein dilatation neurofibromatosis and obstruction of the origin of the large brachiocephalic arteries Notching of the ribs is uncommon in children The lack of rib notching in older individuals usually denotes a mild constriction of the aorta or lack of collateral circulation due to the presence of a patent ductus arteriosus It may also reflect an abnormally low coarctation of the thoracic or abdominal aorta *Unilateral right notching* signifies coarctation proximal to or at the origin of the left subclavian artery In some instances in spite of absence of rib notching a well developed collateral circulation may be demonstrated by angiocardiography

In most instances of coarctation of the aorta angiocardiography is not necessary to prove the presence of this lesion It is however the only method which will clearly outline the length of coarctation *and diameter of the aorta proximal to and* distal to the site of narrowing In the experience of some observers retrograde aortography is preferable to intravenous angiocardiography in the contrast visualization of the aorta

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ELONGATION AND DILATATION

Elongation and Dilatation of the Aorta Acquired lesions of the aorta are largely characterized by structural changes of its wall which are often associated with alterations in the circumference (dilatation) and length (elongation) Such caliber changes makes the recognition of aortic disease amenable to radiologic examination

Under physiologic conditions the elastic elements of the aorta permit volume changes with the inflow of blood from the left ventricle If blood flow and blood pressure are pathologically increased a dynamic dilatation of the aorta may occur which in some instances will lead to irreversible changes In the presence of inflammatory and degenerative processes the elastic elements of the aorta may be damaged to such an extent that very marked caliber changes may develop In addition to increased aortic width and length degenerative calcification of the aortic wall is a significant clue in the radiologic detection

syphilitic aortitis may contribute to the dilatation of the proximal aorta. In patients with aortic stenosis a post-stenotic dilatation of the aorta is frequently observed.

Radiologically elongation and dilatation of the aorta are best demonstrated in the frontal and left anterior oblique projections. Inasmuch as large segments of the aorta cannot be separated from adjacent mediastinal structures, measurements of aortic diameters are subject to many inaccuracies not only in the normal but particularly in patients with chest deformities, pulmonary or pleural disease and diaphragmatic or mediastinal displacement. More precise mensuration of the aortic diameters is only possible *with the use of angiocardiography*. Based on this method Dotter and Steinberg have presented normal values for aortic diameters at various levels. They have shown that in normal subjects aortic width increases with advancing age.

In spite of the well recognized shortcomings of conventional x-ray methods, careful radiographic and fluoroscopic observations of the aorta in various projections allow valuable conclusions as to its true dimensions. This applies especially to the estimation of the diameter of the descending arch where the aortic cross section may be visualized with the help of the esophagogram provided that proper sagittal projections are selected (Kreuzfuchs method). Under pathologic conditions the diameter of the descending aorta is often clearly visible in lateral and oblique projections.

In the frontal view elongation of the aorta is characterized by an increased distance between the right cardiovascular junction and the aortic knob which is displaced upward and to the left towards the level of the clavicles. The descending aorta is seen to bulge widely to the left. In some instances, however, it may be sufficiently tortuous to protrude far into the *right lung field*. In the *left anterior oblique* and lateral projection the aorta is uncoiled and shows a wider than normal swing overlapping the spine posteriorly with formation of a large aortic window.

As the aorta is firmly attached to the base of the heart and diaphragm, pronounced elongation of the aorta may result in bizarre configurations of this blood vessel. With extreme degrees of tortuosity actual kinks of the aorta may be observed which are most commonly found in the region of the ascending aorta, the descend-



Fig. 222. Kinking and dilatation of thoracic aorta. The supradiaphragmatic portion of the descending aorta is situated to the right of the spine simulating a mediastinal mass (arrows).

Fig 220 Elongation of the aorta The bulge of the aorta in the left lung is largely due to elongation as shown in the lateral view The aorta is uncoiled and overlaps the spine There is only mild dilatation present

Fig 221 Elongation and dilatation of the thoracic aorta The aortic arch and descending orta show heavy calcium deposits



Fig 224 Marked kinking of aorta accentuated by kyphosis The descending aorta is angulated in its midportion and bulges into the left lung field (arrows)

only be determined by angiocardiographic procedures which have revealed that the dilatation is greatest in the ascending portion of the aorta and less marked in its distal segment Irregularity of the aortic lumen is also observed on angiocardiography in this condition but it is probably not different from that caused by atheromatous plaques If dilatation of the ascending aorta can be demonstrated and other causes of dilatation such as hypertension and aortic valvular lesions can be excluded syphilitic aortitis should be suspected Early dilatation of the aorta cannot be



Fig 225 Elongation and dilatation of descending aorta The distal esophagus is displaced by the dilated and elongated aorta in the supradiaphragmatic region

Fig 226 Luetic aortitis. In the frontal view delicate calcium deposits are visible in the region of the ascending aorta (arrows)

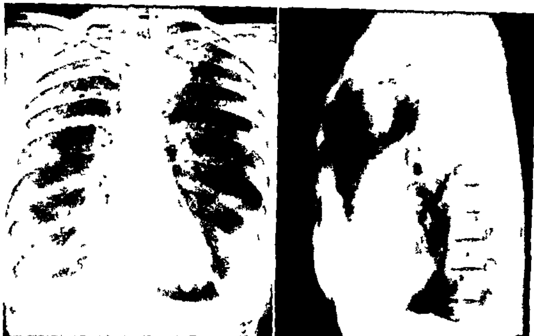


Fig 223 Elongation and tortuosity of thoracic aorta In the mid portion of the descending aorta a slight kinking is visible in the lateral view

ing arch and supradiaphragmatic portion Kinking in the distal arch may lead to formation of a double aortic knob in the frontal projection a configuration which has been described as pseudo coarctation If a localized segment of the aorta protrudes to the right or left of the spine it may simulate the presence of a mediastinal tumor

In the frontal projection a bulge of the ascending aorta to the right beyond a perpendicular tangential line drawn through the right heart border usually indicates aortic dilatation In the left anterior oblique view widening of the ascending aorta may be indicated by a more pronounced forward bulge of this vessel Dilatation of the descending aorta is often indicated by a wide bayonet like kink in the descending aorta just above the level of the diaphragm In the left anterior oblique view limited segments of the aorta are clearly visible against surrounding structures and are subject to direct measurements

In senile ectasia pronounced dilatation and elongation of the aorta is uncommon The caliber changes are usually slight or moderate unless aggravated by other pathologic conditions

Simple atherosclerosis of the aorta does not lead to significant elongation and dilatation of the blood vessel If the disease is more extensive and associated with hypertension syphilis or senile ectasia more pronounced degrees of elongation and dilatation may ensue The calcium plaques of atheromatosis are readily visible on radiologic examination They are most prominent in the region of the aortic knob and descending aorta It is noteworthy that in atheromatosis calcification of the immediate supravalvular region of the aorta is also frequently observed

The predominant involvement of the proximal aorta in luetic processes is of clinical and radiologic diagnostic significance Calcification of the ascending aorta out of proportion to that occurring in the descending aorta makes the presence of a syphilitic infection highly probable It is characteristic of luetic aortitis that dilatation is frequently more pronounced than elongation

The exact size of the blood vessel can



Fig. 229. Dilatation, elongation and kinking of the aorta in a patient with luetic aortitis.



Fig. 230. Tortuosity of innominate artery. The tortuous blood vessel bulges into the right upper lung field. There is associated tortuosity of the aorta (arrows).



Fig. 231. The elongated innominate artery casts a distinct shadow in the superior mediastinum. The outer contour of the trachea remains clearly visible, as the blood vessel is situated anterior to the trachea. A paratracheal mass would obscure the outer contour of the trachea.



Fig. 232. Dilatation and tortuosity of the brachiocephalic arteries in coarctation of the aorta. The dilated vessels are seen as rounded masses in the superior mediastinum (arrows). On operation a marked dilatation of both subclavian arteries and of the left third intercostal artery was noted. Following resection of the coarctation the dilatation of the blood vessels receded. Courtesy of I. A. Mucci, M.D., VA Hospital, Oteen, North Carolina.

Fig 227 Luetic aortitis in a 48 year old female with positive serology Marked calcification of the ascending aorta is visible in both frontal and lateral projections (arrows)



Fig 228 Dilatation and tortuosity of aorta in luetic aortitis The waviness and nodular contour of the aorta simulates the appearance of a *dissecting aneurysm* *The autopsy* disclosed luetic aortitis and no evidence of dissection



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aneurysm or in the thrombotic masses in its lumen

Arteriosclerotic aneurysms of the aorta are less frequently encountered than luetic aneurysms. However, with the diminishing incidence of cardiovascular syphilis and increase of lifespan of the population, arteriosclerotic aneurysms are becoming more prevalent according to recent statistics. These aneurysms are usually smaller and more commonly found in the region of the arch and distal segments of the aorta.

Congenital aneurysms, single or multiple, have been recognized in increasing numbers. Fusiform aneurysms of the sinus of Valsalva are often congenital in nature. In arachnodactyly medial degeneration of the aorta is believed to be an important etiologic factor in the development of this entity. In patients with coarctation of the aorta hypertension is considered to be essential in the formation of sinus of Valsalva aneurysms. Other congenital aneurysms occur in the region of the descending arch of the aorta at the site of entry of the ductus arteriosus.

Of increasing importance is the occurrence of traumatic aneurysm of the aorta which have the descending portion of the arch distal to the origin of the left subclavian artery as site of predilection. This segment of the aorta is relatively fixed by the large vessels and the insertion of the ligamentum arteriosum. Distal to this region the aorta is considerably more flexible and subject to sudden changes of position by violent external trauma. Under these circumstances a shearing off effect may occur at the junction of the two aortic segments incident to a variety of traumatic conditions, particularly automobile accidents with rapid deceleration. Traumatic lacerations of the ascending or distal descending aorta are much less common. If following an aortic tear hemorrhage remains localized or accumulates under the adventitia, a chronic false aneurysm may develop. Generally the chronic traumatic aneurysms have followed a benign course. Conceivably some of the aneurysms of the descending aortic arch which have been designated as congenital in character are actually post traumatic in origin.

Following bacterial infection of the aorta



Fig. 233 Multiple aneurysms of the aortic arch and descending aorta due to syphilis. The aneurysms cause a bilateral mediastinal bulge. The wall of the aneurysms contain calcium deposits.

detected by conventional radiologic methods

Elongation and Dilatation of the Brachiocephalic Arteries Generally those conditions which produce elongation and dilatation of the aorta may also lead to pathologic changes in the width and length of the large vessels Elongation dilatation and tortuosity of the brachiocephalic arteries is commonly referred to as buckling Clinically this process appears to be entirely benign and is of importance only in the differential diagnosis of aneurysm and neoplasm The innominate artery and right subclavian artery are most frequently involved though this process has been noted in the other large vessels arising from the aortic arch The tortuous artery may produce a visible swelling in the supraclavicular region the suprasternal notch and in the soft tissues of the neck In some instances a pulsating mass in the posterolateral pharyngeal wall has been observed On auscultation a bruit may be heard in the region of the buckled blood vessel Buckling of the brachiocephalic arteries is usually encountered in patients with hypertension arteriosclerosis and obesity Striking dilatation of the large vessels has also been observed in patients with coarctation of the aorta

Under normal conditions the innominate artery and large vessels are contained within the mediastinal shadow and do not protrude significantly into the adjacent lung fields However in pronounced buckling a prominence of the large vessel against the adjacent lung fields may be observed in frontal roentgenograms of the chest It is noteworthy that buckling of the large vessels is usually found in association with a tortuous uncoiled aorta In coarctation of the aorta the dilated subclavian arteries are often visible as bulbous or nodular widenings of the superior mediastinum If differential diagnosis between aneurysm and neoplasm is required angiocardiography has to be performed in order to demonstrate the exact dimensions of the involved blood vessels

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ANEURYSMS

The large majority of aneurysms of the aorta and its branches are syphilitic in character Destruction of the muscular and elastic elements of the arterial wall in this infection results not only in diffuse dilatation but may lead to localized bulges particularly along the greater curvature of the blood vessel Multiplicity of syphilitic aneurysms is common and in some instances secondary outpouching of an aneurysm may lead to a multiloculated lesion (daughter aneurysm) Consistent with the prevalence of the luetic infection in the ascending aorta most thoracic aneurysms are found in this region Calcification frequently develops in the wall of the

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mass may be polycyclic in appearance Compression atelectasis of the adjacent pulmonary parenchyma may contribute to a slight irregularity of the profile of the aneurysm

Calcium deposits in the wall of the aneurysm are commonly observed They may consist of delicate curvilinear shadows or irregular plaques not unlike those found in atheromatosis One should keep in mind that calcification similar to that occurring in aneurysms of the aorta may also be found in certain mediastinal neoplasms such as teratomas and thymomas

In the opinion of the authors radiologic determination and evaluation of pulsation of aortic aneurysms is of limited value Though the majority of aneurysms are found to pulsate on fluoroscopy, it appears nearly impossible to differentiate between transmitted and expansile pulsation Many aneurysms lack pulsation due to the accumulation of thrombotic masses or tautness of overlying structures On the other hand striking pulsations are occasionally found in mediastinal neoplasms

Concurrent diffuse dilatation of the aorta in patients with luetic aneurysms is

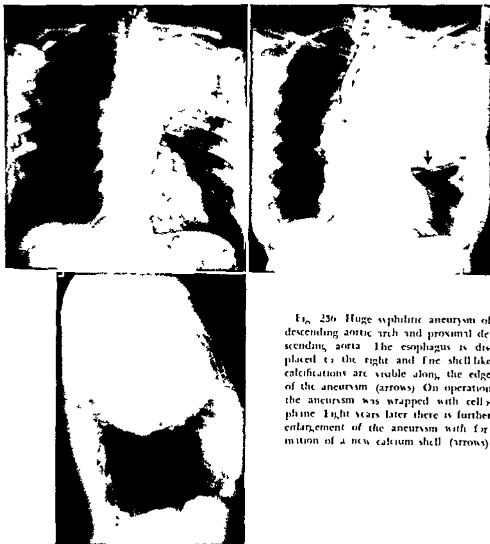


Fig 236 Huge syphilitic aneurysm of descending aortic arch and proximal descending aorta The esophagus is displaced to the right and fine shell like calcifications are visible along the edge of the aneurysm (arrows) On operation the aneurysm was wrapped with cellophane Eight years later there is further enlargement of the aneurysm with formation of a new calcium shell (arrows)



Fig 234 Multiple congenital aneurysms of the distal aortic arch On angiocardiography the aneurysms opacify with contrast medium The diagnosis was confirmed at surgery

a mycotic aneurysm may develop This type of aneurysm is relatively rare and occurs most often in the region of the sinus of Valsalva close to the attachment of the aortic cusps

The following general concepts apply to the **radiologic diagnosis** of aortic aneurysm Any thoracic mass which on radiologic examination cannot be separated from the aorta and large vessels should be regarded as a potential aneurysm Usually the roentgen examination is best carried out by fluoroscopic studies which permit determination of the relationship between the mass and the aorta in various projections

The contour of the thoracic aneurysm forms frequently a smooth arc but with the formation of daughter aneurysms or multiple lesions the edge of such a mediastinal



Fig 235 Syphilitic aneurysms producing atelectasis of the left lower lobe Note the displacement of the esophagus to the right side The calcium rings (arrows) represent 2 aneurysms Surgery revealed that one aneurysm arose from the aortic arch and the other from the left subclavian artery

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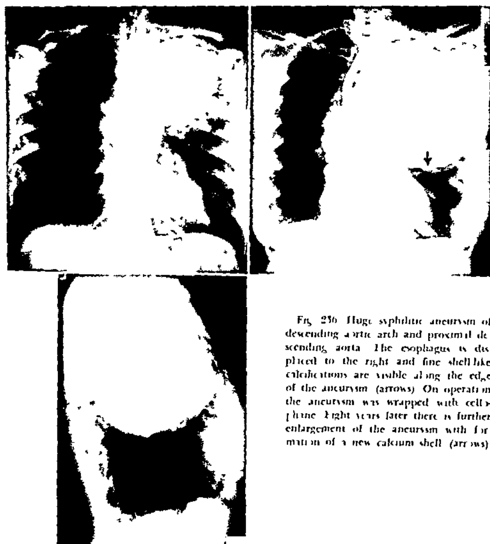


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a very common observation and an important diagnostic aid in the diagnosis of this lesion In the absence of luetic aortic insufficiency the heart in patients with luetic aneurysms may not reveal any pathologic alterations

As the aortic aneurysm expands displacement and compression of adjacent mediastinal structures may bring forth important radiologic changes Compression and obstruction of the trachea and bronchi will lead to the development of obstructive emphysema and subsequent atelectasis In addition obstruction of the superior vena cava or its tributaries may be encountered Incident to compression and stretching of the phrenic and recurrent laryngeal nerves paralysis of diaphragms and vocal cords may occur As the mediastinal contour lines are frequently obscured in partial or complete pulmonary atelectasis diligent search for other radiologic features of tho-

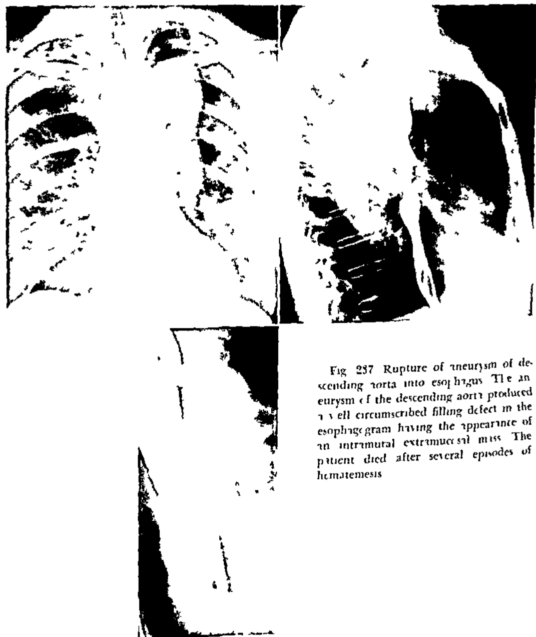


Fig 237 Rupture of aneurysm of descending aorta into esophagus The aneurysm of the descending aorta produced a well circumscribed filling defect in the esophagogram having the appearance of an intramural extramucosal mass The patient died after several episodes of hematemesis





~~C~~alcified aneurysm of ascending ~~a~~neurysm produces a deep smooth ~~th~~e body of the sternum (arrows)

Fig 239 Saccular aneurysm of descending aorta with deep erosion of the adjacent bodies of the eighth and ninth dorsal vertebrae





~~L~~arge aneurysm of descending aortic arch The trachea and esophagus are marked[illegible] the right and anteriorly The left lung shows atelectasis and the left diaphra[illegible] On angiocardiography the aneurysm is opacified (arrows) and a fusiform an[illegible] descending aorta is also outlined

racic aneurysm such as calcification and bone erosion is always indicated On occasions the growth of luetic aneurysms is very protracted, extending over many years Under these circumstances a rearrangement of the peripheral calcifications of the lesion may be observed Rupture of the aneurysm into adjacent mediastinal organs may produce bizarre radiologic changes often caused by mediastinal hematoma retropleural hemorrhage and fistula formation between the aneurysm and the respiratory or gastrointestinal tracts

One of the characteristic features of aortic aneurysm is the erosion of adjacent bony structures Depending upon the location and projection of the aneurysm such parts of the bony thorax as the sternum ribs and spine may be affected Bone erosion is however not a pathognomonic feature of aneurysm as it may be also found in certain mediastinal neoplasms notably lesions of neurogenic origin For the radiologic study of aneurysm fluoroscopy and Bucky roentgenograms of the chest in various projections are often essential Only

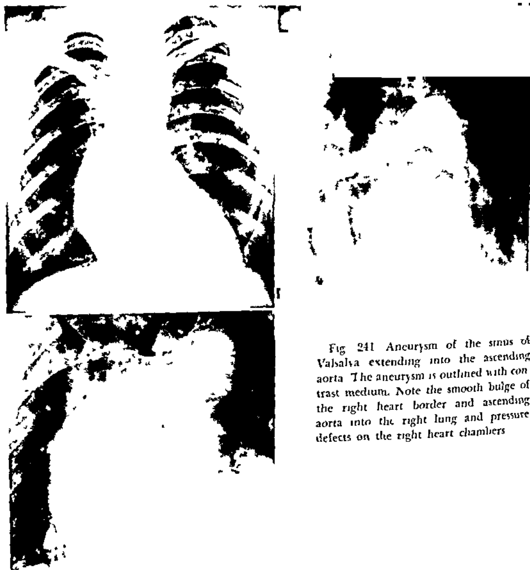


Fig 241 Aneurysm of the sinus of Valsalva extending into the ascending aorta The aneurysm is outlined with contrast medium. Note the smooth bulge of the right heart border and ascending aorta into the right lung and pressure defects on the right heart chambers

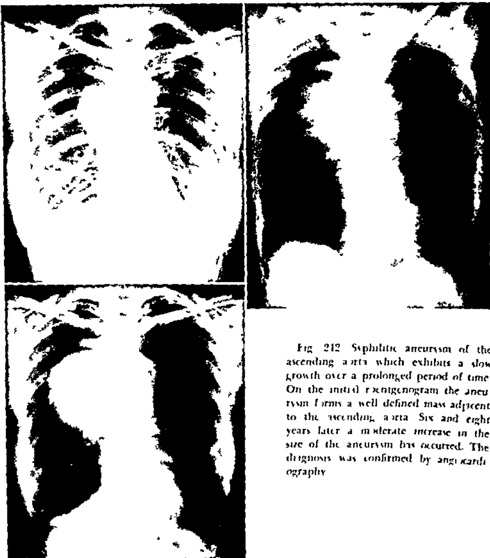


Fig. 212. Syphilitic aneurysm of the ascending aorta which exhibits a slow growth over a prolonged period of time. On the initial roentgenogram the aneurysm forms a well defined mass adjacent to the ascending aorta. Six and eight years later a moderate increase in the size of the aneurysm has occurred. The diagnosis was confirmed by angiocardiography.

In this manner all features contributing to the diagnosis of aneurysm may be recognized. Nevertheless, in some instances angiocardiography is necessary for correct diagnosis and recognition of thoracic aneurysms. There are only very few aneurysms which will fail to show some degree of opacification provided that proper angiocardiographic techniques are selected. This method also serves to delineate exactly the physical dimensions of the aneurysm and aid in a better evaluation of other segments of the diseased aorta. For this reason angiocardiography is often employed in those cases in which surgical resection of an aneurysm is contemplated.

The special **radiologic features** of aneurysms of the aorta and its branches will be discussed in the following order:

(1) Aneurysm of the sinuses of Valsalva
(2) Aneurysm of the ascending aorta
(3) Aneurysm of the aortic arch
(4) Aneurysm of the descending aorta
(5) Aneurysm of the large brachiocephalic arteries
(6) [illegible] neurysm of the [illegible]

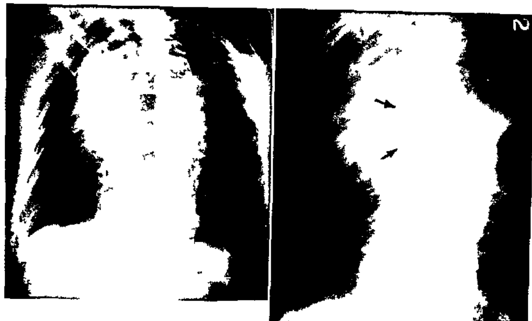


Fig 243 Large syphilitic aneurysm of ascending aorta involving the origin of the large vessels The center of the aneurysm has been opacified on angiocardiography (arrows) The incomplete filling of the aneurysm is the result of blood clot formation

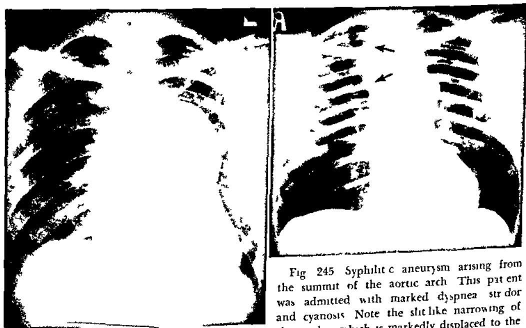


Fig 244 Large syphilitic aneurysm of ascending aorta bulging to the left side

Fig 245 Syphilitic aneurysm arising from the summit of the aortic arch This patient was admitted with marked dyspnea stridor and cyanosis Note the slit like narrowing of the trachea which is markedly displaced to the right (arrows) The autopsy disclosed a saccular aneurysm with erosion of the trachea

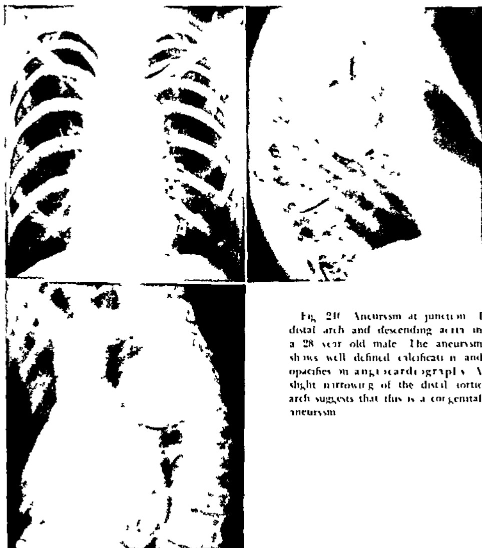


Fig. 210. Aneurysm at junction of distal arch and descending aorta in a 28 year old male. The aneurysm shows well defined calcification and opacifies on angiocardiography. A slight narrowing of the distal aortic arch suggests that this is a congenital aneurysm.



Fig 247 Traumatic aneurysm of aorta A 33 year old patient had a severe chest trauma several years prior to hospital admission The calcified aneurysm (arrows) appears at the junction of the distal aortic arch and descending aorta The aneurysm was surgically removed



Fig 248 Syphilitic aneurysm of descending aorta Note the marked anterior displacement of the esophagus and erosion of the eighth dorsal vertebra (arrows) Subsequent death from rupture into the left pleural space

Aneurysm of the aortic sinuses (sinuses of Valsalva) may be fusiform or saccular in shape Most of these aneurysms are syphilitic or mycotic in origin but recently an increasing number of these lesions have been described in association with coarctation of the aorta arachnodactyly and subaortic stenosis

As the aortic sinuses are largely surrounded by other cardiac structures the clinical symptomatology as well as radiologic findings in these patients may be very bizarre Due to the close anatomic relationship of sinus aneurysms rupture into adjacent structures such as the pulmonary conus the pulmonary artery and the atria may lead to unusual derangements of cardiovascular dynamics An aneurysm rupturing into the pulmonary artery may lead to considerable increase of pulmonary artery pulsation and pulmonary congestion

Many aneurysms of the aortic sinuses are contained within the cardiac mass and may not reveal any characteristic changes on conventional radiologic procedures If the aneurysm is sufficiently large it may project from the confines of the cardiac structure and participate in the formation of the right or left heart border Calcification of these aneurysms is occasionally recognized and may be diagnostic even in the absence of extracardiac mass formation With rupture of an aortic sinus aneurysm into the pulmonary artery an increased pulmonary blood flow is reflected by unusual pulsations of the pulmonary artery In general angiocardiography is the only procedure which determines the exact location and extent of these aneurysms

Aneurysms of the ascending aorta are usually syphilitic in character and represent the most common group of all aneurysms Other rare causes of aneurysm of the ascending aorta are atherosclerosis trauma and congenital heart disease Aneurysms of the proximal aorta may be

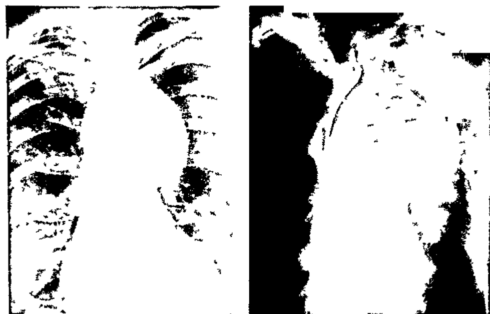


Fig 219 Aneurysm of the aorta with associated esophageal fistula The initial examination disclosed a moderate enlargement of the aortic arch and compression and displacement of the trachea and esophagus The lesion was wrapped with cellophane and patient was discharged improved One month later following several days of hematemesis a re-examination disclosed enlargement of the aneurysm and extravasation of contrast medium from the esophagus into the peripheral portion of the mass Death occurred from hemorrhage a short time la

fusiform or saccular in shape and may assume occasionally huge proportions Small aneurysms if confined to the root of the aorta or directed posteriorly may escape radiologic detection If the aneurysm bulges to the right it is usually best demarcated in the left anterior oblique position Under these circumstances the trachea as well as the esophagus are displaced to the left side and posteriorly More rarely expansion of the aneurysm to the left side takes place where it may dislodge the pulmonary artery and appear as a well circumscribed shadow overlying the waist line of the heart imitating the pulmonary trunk Aneurysms of the ascending aorta may cause erosion of the rib cage and sternum a phenomenon of considerable clinical importance as it is rarely observed in mediastinal neoplasms Compression of the trachea and right main stem bronchus may lead to broncho stenosis with atelectasis Occasionally paralysis of the right diaphragm is observed as the result of injury of the right phrenic nerve

Most *aneurysms of the aortic arch* are syphilitic though in the distal segments of the aortic arch an increasing number of arteriosclerotic congenital and traumatic aneurysms are now being recognized Aneurysm of the aortic arch is often called the aneurysm of symptoms Though small the close proximity of adjacent organs and the limited space of the thoracic aperture may produce pronounced clinical symptoms out of proportion to the size of the aneurysmal sac

In most instances the trachea is displaced to the right and may be compressed to a slit like lumen The tracheal displacement is often well circumscribed and located at the level of the aneurysmal sac whereas tracheal displacement in patients with



Fig 250 Syphilitic aneurysm of innominate artery A 52 year old patient developed a pulsating suprasternal mass which displaced on radiologic examination the esophagus and trachea to the left side and posteriorly On operation an aneurysm of the innominate artery was found which involved the aortic arch

Fig 251 Innominate artery aneurysm probably traumatic *This patient* sustained a severe chest trauma in a car accident four years prior *to admission* The aneurysm was successfully resected Courtesy of A G Singer M D Toccoa Georgia



Fig 252 Traumatic aneurysm of left subclavian artery This patient had a severe crushing injury of the chest resulting in mediastinal hematoma which gradually subsided Seven years later a pulsating mass developed in the left supra-aortic region (arrows) Tomograms demonstrated calcium deposits in the base of the mass The aneurysm was removed surgically



Fig 253 Dissecting aneurysm of aorta The roentgenogram prior to onset of symptoms shows a tortuous aorta and left ventricular enlargement Striking widening and irregularity of the aortic contour developed following onset of severe chest pain radiating to the dorsal spine The autopsy disclosed dissecting aneurysm of the aortic arch and descending aorta



Fig 254 Dissecting aneurysm of aorta in a 42 year old patient with hypertension Note the dilatation of the descending aorta and wavy outer contour of the blood vessel The aneurysm ruptured into the left retropleural space

goiter usually extends to the region of the neck Also on swallowing upward motion of the aneurysm is much less marked than that of a goiter which is more firmly attached to the trachea

If the aneurysm originates from the most anterior portion of the aortic arch the trachea may be displaced posteriorly otherwise the trachea and esophagus are dislodged anteriorly and to the right Occasionally an aneurysm may dissect into the space between trachea and esophagus separating the two organs Depending upon location of the aneurysm the sternum as well as the proximal vertebral column may show signs of bone erosion Such bone erosion is usually best demonstrated in the right anterior oblique position

Compression of the trachea and left main bronchus by aneurysms of the aortic arch may lead to partial or total atelectasis of the left lung In many instances paralysis of the left phrenic nerve results in elevation of the left diaphragm and paradoxical motion

Aneurysms of the descending aorta assume in many instances huge dimensions without producing significant clinical symptoms If small aneurysms of this type may be entirely obscured by the cardiac shadow in the frontal projection These aneurysms usually deviate the esophagus to the right and anteriorly

Aneurysms of the descending aorta are best demonstrated in both oblique projections with the use of grid roentgenograms which may also delineate erosion of the left antero lateral aspect of the vertebral column As the esophagus is wedged between the aneurysm and the heart esophageal erosion and fistula formation has been observed in this region resulting in profuse esophageal hemorrhage

Similar to aneurysms of the aorta *aneurysms of the large vessels* may be syphilitic arteriosclerotic or traumatic in origin These aneurysms may be fusiform or saccular in shape occasionally assuming huge dimensions Syphilis is presently the most common cause of innominate artery aneurysm It is likely that in the future as syphilis becomes less common arteriosclerotic and traumatic aneurysms will play a more important role

If intrathoracic the aneurysm forms a mass in the supra aortic portion of the mediastinum which may or may not pulsate on fluoroscopic examination Shell like calcium deposits in the aneurysm are demonstrated to advantage on grid roentgenograms Erosion of the ribs and sternum are common in syphilitic aneurysms of the innominate artery Downward displacement of the aortic knob is often encountered and lateral and posterior deviation of the trachea and esophagus by the aneurysm should be searched for The aneurysm may occasionally move with swallowing but this motion is usually not as striking as in thyroid tumors Angiocardiography is necessary for exact delineation of the aneurysm and differentiation of this mass from simple elongation and dilatation of the affected blood vessel

The development of *dissecting aneurysm* is closely linked with a degenerative proc

ess of the media consisting of necrosis and cyst formation. Hypertension is considered to be an important but not essential contributing factor.

Hemorrhage may develop at the site of cystic media degeneration apparently as the result of the rupture of a vas vasorum. As the hemorrhage distends the medial layers of the aorta a tear of the intima may occur thus opening the space of dissection to the blood flow and blood pressure of the aorta. This tear is usually transverse in position and most often found in the supravalvular area or close to the ostia of the brachiocephalic arteries. Splitting of the aortic wall may be extensive extending throughout the entire length of the blood vessel. Re-rupture through the intima may take place at some distance and may in some cases lead to spontaneous recovery. A tear of the adventitia will lead to rupture into the body cavities particularly the pericardium. A frequent and grave complication is occlusion of the mouth of the coronary arteries and other aortic branches by the dissecting process.

Unfortunately the serious clinical condition of the patient makes a satisfactory examination often impossible. A common finding is the widening of the aortic shadow in the frontal projection. If there is extension of the process to the brachiocephalic vessels a diffuse widening of the supra-aortic mediastinal shadow may be found. The widening of the aortic shadow in itself cannot be considered as pathognomonic as it may be similar to that of other types of aortic dilatation. If rapid changes in the aortic contour can be established by comparison with previous roentgenograms the diagnosis of dissection appears however assured. The outer contour of the aorta may show a wavy angular and somewhat irregular appearance as the result of wall distention. A similar appearance of the outer contour of the aorta may also be observed in luetic aortitis.

Sometimes the radiologic features of the complications may interfere with recognition of the aneurysm. Hemorrhage into the mediastinum or pleural fluid collection are commonly observed.

Recently angiocardiographic procedures have been employed in the diagnosis of dissecting aneurysm. With this method the site of the dissection may be detected and the extent of the involvement demonstrated by encroachment of the aortic lumen (Golden and Weens). Angiocardiographic studies in this condition have now



Fig 2[illegible] Dissecting aneurysm of the aorta. The aorta is diffusely widened and reveals a wavy nodular outer contour.



Fig 256 *(See next page)*

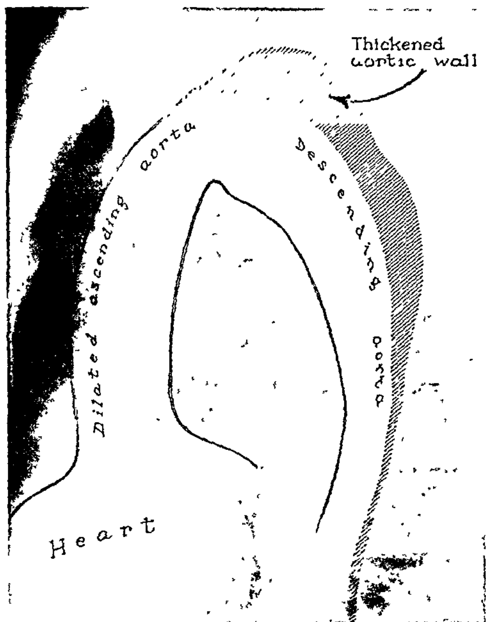


Fig 56 Dissecting aneurysm demonstrated with angiocardiography The angiocardiogram outlines the aortic lumen which is narrowed in the descending portion of the aorta Note the distension of the aortic wall along the greater curvature From Golden A and Weens H S The diagnosis of dissecting aneurysm of the aorta by angiocardiography *Am Heart J* 37 114 1949



Fig 258 Hypoplasia of the right pulmonary artery The right lung is underdeveloped and the left lung reveals compensatory enlargement On angiocardiography an abnormally small right pulmonary artery was noted A small artery arising from the descending aorta supplied the right lower lobe There was associated malformation of the right bronchial tree

anomaly has been observed on the right side as an isolated malformation On the left the abnormality has usually been recognized in conjunction with tetralogy of Fallot

A rare but clinically important anomaly is abnormal course of the left branch of the pulmonary artery In this malformation the left pulmonary artery runs to the right side and surrounds the distal trachea before proceeding to the left hilum Respiratory distress has been a characteristic clinical feature in the observed cases In its abnormal course the left pulmonary artery passes between the trachea and esophagus which show compression defects on radiologic examination with contrast medium

Dilatation of the pulmonary artery is encountered in many clinical entities most of which are characterized by pathologic alterations of blood flow and blood pressure of the pulmonary circulation

Dilatation of the pulmonary artery and its main branches develops in cardiac malformations with increased pulmonary blood flow notably in interventricular and interatrial septal defects patent ductus arteriosus and abnormal pulmonary venous return Among the acquired cardiac conditions thyrotoxicosis and beri beri heart



Fig 259 Two cases of pulmonary artery dilatation in patients with intracardiac shunt The pulmonary artery bulges into the adjacent left lung field (arrows)



Fig 260 Post stenotic dilatation of the pulmonary artery in a patient with pulmonary stenosis The dilatation of the pulmonary artery is confined to the post valvular segment of the pulmonary trunk (arrows)

Fig (... mum l ... monar



Fig 262 Aneurysm of main trunk of pulmonary artery and its main branches Marked calcification of the arterial walls is visible (arrows) The autopsy disclosed a patent ductus arteriosus measuring 1 cm in diameter Intracardiac shunts were suspected during life but not found on autopsy



Fig 263 Aneurysm of pulmonary artery in a patient with a large patent ductus arteriosus The aneurysm involves the main pulmonary artery and both branches but was most pronounced on the right side

disease deserve foremost consideration In some instances turbulence of pulmonary blood flow such as seen in pulmonary stenosis may result in a definite ectasia of the blood vessel In pulmonary stenosis the arterial dilatation is confined to the post valvular segment of the pulmonary trunk

Pathologic alterations of pulmonary arterial pressure may occur in a large variety of clinical situations leading to dilatation and elongation of the pulmonary artery In addition to various types of congenital heart disease acquired cardiac conditions such as mitral stenosis and pulmonary valvular disease may produce this phenomenon Dilatation of the pulmonary artery is also observed in pulmonary arteriosclerosis pulmonary embolism and in pulmonary and pleural diseases leading to increased resistance in the lesser circulation

Intrinsic changes of the arterial wall on a degenerative or inflammatory basis weaken the blood vessel resulting in localized or fusiform ectasia Foremost among these conditions are atheromatosis bacterial infection and rarely syphilis In many instances these pathologic changes are found in conjunction with alteration of pulmonary blood flow and pressure

Radiologically precise determination of the dimensions of the pulmonary artery in its main trunk and principal branches within the mediastinum is not feasible by conventional roentgen procedures Direct measurement of the pulmonary artery depends entirely upon angiocardiographic methods Nevertheless observations of the mediastinum in frontal oblique or lateral projections on fluoroscopy and with the aid of various radiographic techniques such as tomography will frequently offer valuable clues as to the presence of pulmonary artery dilatation

Under normal conditions the mid portion of the left heart border is formed by the left lateral contour of the main trunk of the pulmonary artery sometimes in conjunction with its left descending branch Elongation and dilatation of the pulmonary artery will cause an unusual promi



Fig 264 Calcification of obliterated ductus arteriosus demonstrated on lateral tomogram (arrows) This patient had coarctation of the aorta On operation the calcification of the obliterated ductus was seen to extend into the wall of the aorta

nence or bulge of this segment. In the right anterior oblique projection the anterior border of the pulmonary artery may be recognized against the adjacent lung field just above the level of the pulmonary valve. A forward bulge of this segment usually denotes dilatation of the pulmonary artery. The dilated blood vessel will also displace the esophagus posteriorly by pressure on the adjacent left main bronchus and tracheal bifurcation. Increased pulsation of the dilated pulmonary artery is often better correlated with increased bloodflow than with increased arterial pressure. If atheromatosis is severe, calcification of the main trunk and branches of the pulmonary artery is occasionally observed.

Pulmonary artery aneurysms may be fusiform or saccular in character. In the large majority of cases the main trunk with or without the principal branches is involved. Rarely the right or left pulmonary artery branch is affected exclusively. The separation of dilatation of the pulmonary artery from true aneurysm is difficult. In a strict sense aneurysm should only be considered if in addition to blood vessel dilatation pathologic alteration of one or more layers of the arterial wall has taken place. Even then dilatation of the pulmonary artery with concurrent atheromatosis can hardly be distinguished from what may be defined as a fusiform aneurysm. If the aneurysm is saccular, a clearer definition of the lesion is usually possible.

Based on necropsy observation in two extensive summarizing reports by Boyd and McGavack and Deterling and Clagett syphilis could be considered as the principal etiologic factor in approximately one third of the reported cases. In nearly half of the necropsies the aneurysms appear to have developed on a congenital basis as the result of associated congenital cardiovascular malformations such as a patent ductus arteriosus. In the remaining instances atheromatosis or mycotic degeneration of the pulmonary artery was found. Nevertheless there are instances in which the aneurysmal dilatation of the pulmonary artery cannot be satisfactorily explained.

One should realize that these statistics



Fig. [illegible]. Large patent ductus arteriosus demonstrated by retrograde aortography. The ductus is indicated by arrows. Its size approaches that of the descending aorta.

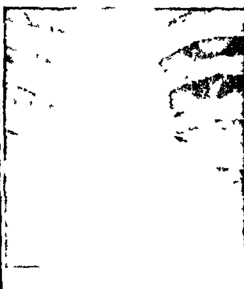


Fig. [illegible]. Diverticulum-like dilatation of the aorta at the site of the entry of a patent ductus arteriosus. The arrows indicate the [illegible] bulg[illegible] the aorta.



Fig 267 Aneurysm of the patent ductus arteriosus The aneurysm causes a widening of the mediastinal shadow Two delicate curvilinear calcifications were noted in this region (arrows)

are based on the evaluation of necropsies and do not necessarily reflect the incidence in clinical practice Thus syphilitic aneurysm of the pulmonary artery appears decidedly rare even in those institutions in which syphilitic aneurysm of the aorta is commonly observed On the other hand due to the interest in the study of congenital heart disease aneurysmal dilatation of the pulmonary artery and its branches is often encountered in the clinical and radiologic studies of various types of congenital malformations

On radiologic examination the aneurysm of pulmonary artery may cause unusual bulge into the adjacent left lung field which is frequently best demonstrated in the oblique projections Calcification develops in some instances and is more clearly recognized by Bucky roentgenograms The true extent of the dilatation is usually shown to advantage by angiocardiographic procedures In some instances the differentiation of pulmonary artery aneurysm from aortic aneurysm or aneurysm of a patent ductus arteriosus may prove to be difficult without resort to specialized radiologic methods Rupture of the aneurysm into adjacent structures as such in the aorta has been reported

The *ductus arteriosus* presents a normal vascular channel between the pulmonary artery and aorta during fetal and early post natal life This blood vessel extends from the descending aorta to the left pulmonary artery just distal to the bifurcation of the main trunk The ductus arteriosus usually obliterates within a few weeks after birth to form the ligamentum arteriosum In some instances this orderly obliteration may lead to the calcification of the ductus close to its aortic entry which may be recognized on radiologic examination in the frontal oblique and lateral projections

Persistent ductus arteriosus may occur as an isolated anomaly or in conjunction with various other cardiovascular malformations A detailed description of the clinical and *radiologic manifestations of patent ductus* arteriosus would be beyond the scope of this monograph The ductus arteriosus is sometimes clearly visible in frontal roentgenograms as a small contour shadow within the confines of the mediastinum above the pulmonary artery segment of the left heart border The pulmonary artery segment in these patients is usually prominent and somewhat more elevated than in other types of pulmonary artery dilatation The ductus itself may be demonstrated by angiocardiographic procedures in oblique and lateral projections preferably by retrograde aortography Increased pulsation of the pulmonary artery segment of the left heart border *in conjunction with prominence* and increased pulsations of the aorta reflects the presence of the abnormal aorto-pulmonary communication

At the level of the entry of the patent ductus arteriosus into the aorta a slight diverticulum like dilatation of this blood vessel is often visible on angiocardiographic



Fig 268 Aneurysm of the ductus arteriosus The frontal roentgenogram shows a bulging mass in the region of the pulmonary artery segment A retrograde aortogram demonstrates an aneurysm of the ductus arteriosus The stump of the ductus filled with contrast medium (arrows) At surgery an aneurysm of the ductus arteriosus was found which was friable and contained organized thrombus Courtesy of W Molnar M D Columbus Ohio

examinations though this is not pathognomonic for this entity Other radiologic features observed in patent ductus arteriosus are left atrial and left ventricular enlargement

Aneurysms of the patent ductus arteriosus are rather rare and may develop on a mycotic or congenital basis The large majority of aneurysms of this type have been observed as incidental necropsy findings in infants often in association with infection Of greater importance is the development of a traumatic aneurysm as a complication of surgical procedures for the ligation of the ductus arteriosus Radiologically these lesions may form a mass protruding into the adjacent lung field above the level of the left hilum Differentiation of the aneurysm from other aneurysms and masses in this region requires frequently specialized radiographic procedures Calcification is observed in some of these lesions and represents a valuable diagnostic feature if the aneurysm is small and obscured within the confines of the mediastinum

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Chapter 28

LESIONS OF THE SUPERIOR VENA CAVA AND ITS TRIBUTARIES

In this chapter congenital anomalies and acquired pathologic changes of the superior vena cava and its tributaries are described

Congenital Anomalies of the Superior Vena Cava and Its Tributaries The anatomic variations of the thoracic venous system are so numerous that they would defy detailed description within the scope of this book Only the most commonly encountered or clinically important conditions shall be considered here Some of these venous changes may cause significant variations of the mediastinal contour It is therefore proper to consider them in the differential diagnosis of mediastinal disease Several of the mediastinal venous abnormalities are closely associated with various types of congenital heart disease making such changes of considerable clinical importance in the diagnosis and therapy of cardiac malformations

A brief review of the developmental anatomy of the thoracic veins will be presented as it may aid in the understanding of these pathologic changes A study of the developmental features of the thoracic venous system may also facilitate the proper recognition and classification of those abnormalities which are not described here

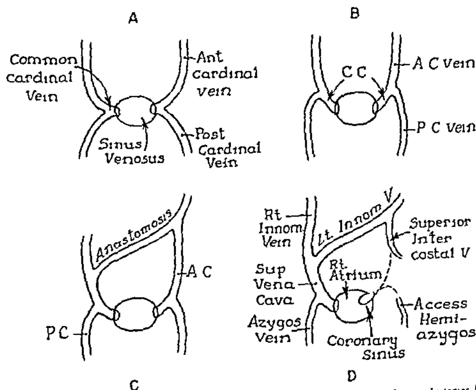


Fig 269 Diagram showing the development of the superior vena cava and its tributaries from the cardinal vein system

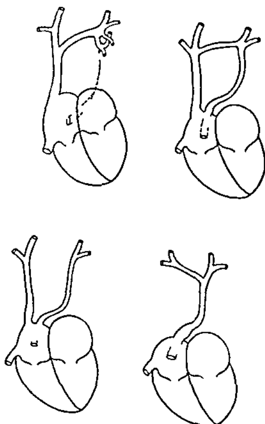


Fig 270 The normal superior vena cava system and some of the more commonly observed anomalies due to persistence of segments of the cardinal veins Note that the persistent left superior vena cava empties into the coronary sinus

Development of the Superior Vena Cava System In the early embryo most of the blood of the body is returned to the heart through the cardinal venous system The anterior cardinal veins collect the blood from the head end of the fetus whereas the posterior cardinal veins gather blood from the lower portion of the trunk These paired venous structures join to form the short common cardinal veins (ducts of Cuvier) which enter the venous sinus of the heart

Further development of the mediastinal venous system takes place by such changes as shifts in position and direction second ary anastomoses atrophy and replacement of existing venous plexus and channels As the fetus grows an important anastomosis between the two anterior cardinal veins is established This channel is the precursor of the left innominate vein and deviates blood from the left head end of the em bryo towards the right anterior and com mon cardinal veins which in turn are dif ferentiated into the superior vena cava system As this development takes place the left anterior and left common cardinal veins undergo atrophy but smaller seg ments of these vessels participate in the formation of the coronary sinus and ob lique vein of the left atrium

If such an orderly obliteration of the left anterior and common cardinal vein system is disturbed larger or smaller parts of these vessels remain active leading to partial or complete persistence of a left superior vena cava system Winter has pre sented an extensive description of the very numerous variations which have been re corded in the literature

Development of Pulmonary Veins In early fetal development the primordia of the lower respiratory tract are derived from the foregut Initially therefore the respira tory tract shares a common blood supply with the circulation of the intestinal vis cera During early developmental phases both foregut and lungs are enveloped in a common capillary network the splanchnic plexus which drains into the cardinal vein system as well as several other visceral veins entering the common cardiac atrium

It is noteworthy that at this stage of development the lungs do not possess a direct venous channel to the heart Such direct venous drainage develops only later by formation of a common pulmonary vein which connects the capillary circulation of the lung with the left atrium This pul monary vein in turn subdivides into sev eral branches which are then incorporated into the left atrium

Failure of the pulmonary veins to prop erly develop results in faulty venous com munication of lungs and heart and may enhance partial or total abnormal pul monary venous drainage into the systemic

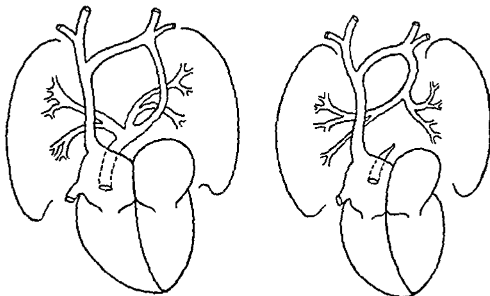


Fig 271 Diagram showing faulty venous connection of the lungs The abnormal pulmonary venous connection enters the left superior vena cava system

veins The abnormal pulmonary venous connection is under these circumstances frequently associated with persistence of the left superior vena cava system

Persistent Left Superior Vena Cava A persistent left superior vena cava is most often encountered as an isolated abnormality If the superior vena cava on the right side is normally developed complete reduplication of the superior vena cava system will ensue In many instances the two vessels are connected by the left innominate vein As the left superior vena cava is derived embryologically from the left cardinal system it usually drains into the coronary sinus which will not cause any alteration of cardiovascular dynamics



Fig 272 Reduplication of superior vena cava system The left superior vena cava and right superior vena cava are connected by a venous channel

In the large majority of cases it is not possible to diagnose a persistent left superior vena cava on frontal roentgenograms Occasionally however, the abnormal vein may cast a distinct shadow of the superior mediastinum or straighten the contour of the left mediastinal border In other cases the vein is shown by a faint bandlike density which gradually blends with the mediastinal soft tissues Generally however accurate information has to be obtained by contrast visualization or cardiac catheterization In both the frontal and lateral projections venographs will demonstrate the entry of the persistent left superior vena cava into the coronary sinus In patients with congenital heart disease persistence of the left superior vena cava is frequently an associated anomaly Under these conditions the abnormal vein may drain into the left atrium and septal defects are commonly observed

Abnormal Pulmonary Venous Return Of considerable importance is the syndrome of abnormal drainage of the pulmonary veins into a persistent left superior



Fig 273 Persistent left superior vena cava in a patient with congenital heart disease The abnormal vessel is seen as a faint shadow in the superior mediastinum on the left side (arrows) The left superior vena cava enters the coronary sinus



Fig 274 Abnormal drainage of pulmonary veins into the persistent left superior vena cava The frontal roentgenogram shows the typical figure of 8 configuration The left superior vena cava serves as a collecting vessel for the pulmonary veins Both venae cavae are enlarged and showed pulsation on fluoroscopy There is marked pulmonary hypervascularity reflecting the increased pulmonary blood flow

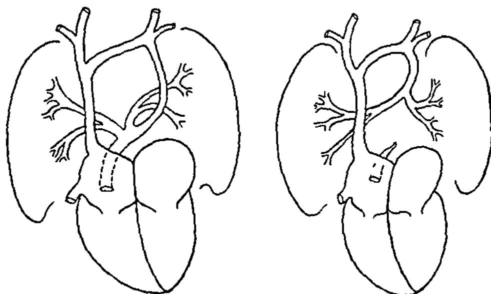


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Fig 273 Persistent *left superior* vena cava in a *patient* with congenital heart disease The *abnormal* vessel is seen as a faint shadow *in the superior* mediastinum on the left side (arrows) *The left* superior vena cava enters the coronary sinus



Fig 274 Abnormal drainage of pulmonary *veins into the* persistent left *superior* vena cava The *frontal* roentgenogram *shows* the typical figure of 8 *configuration* The left superior vena cava serves as a *collecting* vessel for the pulmonary veins Both venae *cavae* are enlarged and showed pulsation on *fluoroscopy* There is marked pulmonary *hypervascularity* the increased pulmonary blood *flow*

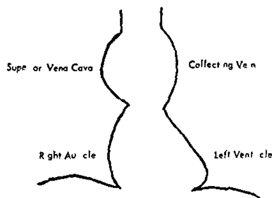


Fig 275 Diagram showing the characteristic deformity of the mediastinum in abnormal pulmonary venous return This deformity has been described as figure of 8 formation mediastinal moustache or cottage loaf shape From Leigh *et al* Venous aneurysms of the mediastinum *Radiology* *63* 696 1954

vena cava as this malformation may cause changes in the configuration of the mediastinum which are highly characteristic and require attention in the differential diagnosis of mediastinal masses In this anomaly the persistent left superior vena cava functions as a collecting vessel for the pulmonary veins

On radiologic examination the distended vein may be found to pulsate and bulge into the left upper lung field The vessel usually drains into the normally developed superior vena cava which is also dilated protruding distinctly into the right upper thorax This bilateral venous bulge above the cardiac shadow is commonly referred to as the figure of eight formation mediastinal moustache or cottage loaf shape In the absence of pulmonary stenosis hypervascularity of the lungs reflects the increased blood flow through the lesser circulation In some instances the venous bulge may be more pronounced on either side resulting in an asymmetric enlargement of the mediastinum

Hypoplasia or Aplasia of Inferior Vena Cava with Azygos Drainage In rare instances congenital underdevelopment or absence of the inferior vena cava will result in a compensatory overdevelopment of the azygos system Under these circumstances the blood from the lower part of the body is channeled to the superior vena cava through a dilated azygos or hemiazygos venous system Though this venous abnormality may exist as an isolated malformation it is most commonly detected with other cardiac malformations especially cor biloculare

The arch of the azygos vein appears as a rounded density in the superior mediastinum which may be mistaken for a mediastinal mass Careful radiologic studies will permit a complete tracing of the entire vessel which may imitate the shadow of an abnormally placed aorta Though this vascular abnormality may be suspected on conventional roentgen procedures angiography is required for exact demonstration of this vessel

Isolated Aneurysmal Dilatation of the Superior Vena Cava An unusual case of isolated enlargement of the superior vena cava and a smaller portion of the azygos vein and internal mammary vein has been described by Leigh and his associates In this instance the superior vena cava apparently on a developmental basis was



Fig 276 Angiocardiogram in a patient with total abnormal pulmonary venous return The contrast medium in the superior vena cava is markedly diluted by the non-opacified blood of the systemic and pulmonary circulation From Leigh *et al* Venous aneurysms of the mediastinum *Radiology* *63* 696 1954

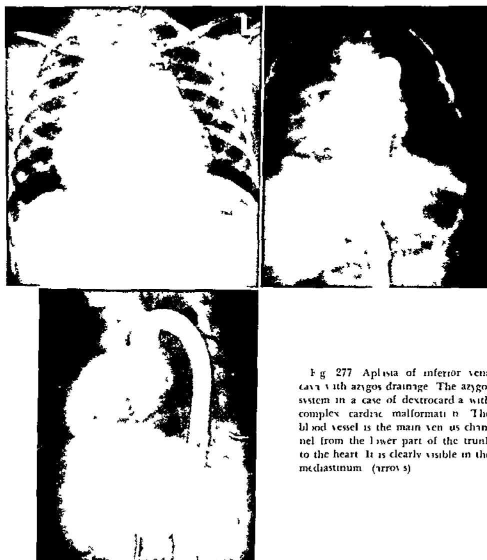


Fig 277 Aplasia of inferior vena cava with azygos drainage The azygos system in a case of dextrocardia with complex cardiac malformation The blood vessel is the main venous channel from the lower part of the trunk to the heart It is clearly visible in the mediastinum (arrows)

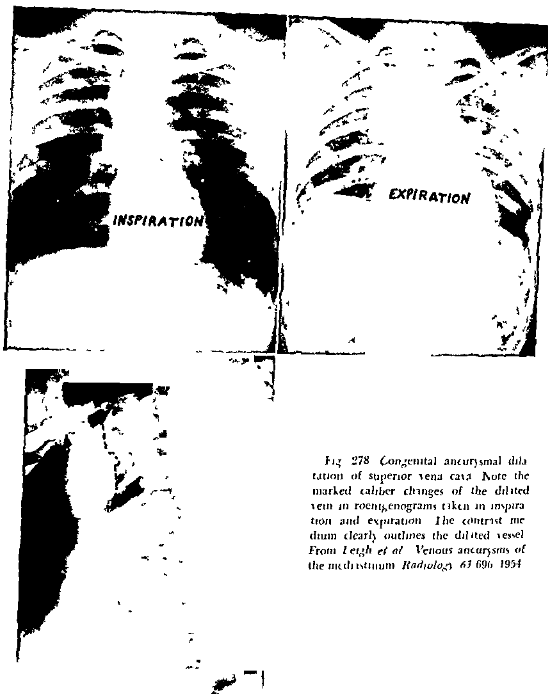


Fig 278 Congenital aneurysmal dilatation of superior vena cava Note the marked caliber changes of the dilated vein in roentgenograms taken in inspiration and expiration The contrast medium clearly outlines the dilated vessel From *Leigh et al* Venous aneurysms of the mediastinum *Radiology* 63 696 1954



Fig 279 The appearance of the normal azygos vein in supine and erect position (arrows) In the erect position the blood vessel is nearly collapsed but it is clearly visible in the supine position as an almond shaped density These changes in caliber differentiate the vein from an enlarged lymph node The mass in the left superior mediastinum consists of enlarged tuberculous lymph nodes

found to be hugely enlarged On surgical exploration there were many small grape like projections along its anterolateral surface

On radiologic examination the dilated blood vessel showed pulsations of very wide amplitude There were also considerable changes in the shape of the dilated vein in roentgenograms taken on deep inspiration and expiration as well as on changes in body position The dilated blood vessel could be readily demonstrated on contrast visualization of the superior vena cava

Acquired Dilatation of the Superior Vena Cava and Its Tributaries Acquired dilatation of the superior vena cava and its tributaries may be found in the presence of elevated venous pressure and increased blood flow Only the more important clinical entities associated with these pathologic changes shall be described here

Venous Dilatation in Heart Disease Dilatation of the large veins of the mediastinum may be noted in congestive heart failure constrictive pericarditis pericardial effusion and tricuspid insufficiency Radiologically enlargement of the venous system will manifest itself in widening of the superior mediastinum This observation is usually of little diagnostic significance in those patients with congestive heart failure in whom such features as diaphragmatic elevation soft tissue edema and pleural fluid collection will often broaden the mediastinal shadow

From the radiologic standpoint changes in the caliber of the azygos vein as it arches over the right main bronchus are often more useful in the recognition of venous engorgement Normally the azygos vein rests in the angle between the trachea and right main bronchus The cross section of this vessel in the anteroposterior projection is rounded ovoid or almond shaped It is noteworthy that the diameter of this blood vessel may change appreciably in the Valsalva or Mueller experiments

Variations in the diameter of the azygos



Fig 280 Rotation and displacement of the mediastinum to the left side reveals a large segment of a normal azygos arch (tomogram) The blood vessel is indicated by arrows

vein may also be affected by changes in position and respiration The lower venous



Fig 281 Azygos vein dilatation in congestive heart failure The dilated azygos vein (arrows) is seen as a round density in the superior mediastinum

pressure in the erect position will be reflected in partial collapse of this blood vessel These physiologic changes in the diameter of the azygos vein may aid considerably in its differentiation from an enlarged lymph node or small tumor

Under normal conditions the diameter of the azygos vein in upright chest roentgenograms rarely measures more than 6 mm in diameter In patients with elevated



Fig 282 Obstruction of the left innominate vein in a patient with a saccular aneurysm of the innominate artery The left innominate vein does not fill with contrast medium Numerous collateral veins empty into the superior vena cava which is displaced to the right side

venous pressure the caliber of this vessel may considerably exceed this value (Fleischner and Udis)

Obstruction of the Superior Vena Cava and Its Tributaries Obstruction of the superior vena cava and its tributaries has been observed in a large variety of clinical entities Most often the obstruction will be found to be due to neoplastic disease aneurysm chronic fibrous mediastinitis and localized phlebitis with thrombus formation Depending upon the site of the obstruction an extensive collateral circula

tion in the mediastinum and thorax will develop in which the azygos system the internal mammary veins the vertebral plexus and lateral thoracic veins participate

On radiologic examination the superior mediastinum is found to be markedly widened by the dilated collateral channels and the veins proximal to the area of obstruction Usually however the underlying cause of obstruction such as tumor or aneurysm are the predominant radiologic features If the obstruction is below the entrance of the azygos vein this vessel may deviate blood flow to the inferior vena cava with little dilatation of the other venous structures of the mediastinum

Obstruction of the Inferior Vena Cava and Portal Vein System As the result of obstruction of the inferior vena cava or of the portal vein and its branches an extensive collateral venous circulation may develop in the mediastinum Though the development of esophageal varicosities is well recognized it is less commonly emphasized that other venous channels of the mediastinum may dilate to such an extent that they may simulate the appearance of mediastinal masses Dilatation of the azygos and hemiazygos system in patients with portal vein hypertension or splenic vein thrombosis has been observed which was sufficiently pronounced to pose a problem in radiologic differential diagnosis of mediastinal tumors

In obstruction of the inferior vena cava distinct dilatation of the azygos vein may become visible on radiologic examination This may be particularly pronounced in the presence of elevated venous pressure

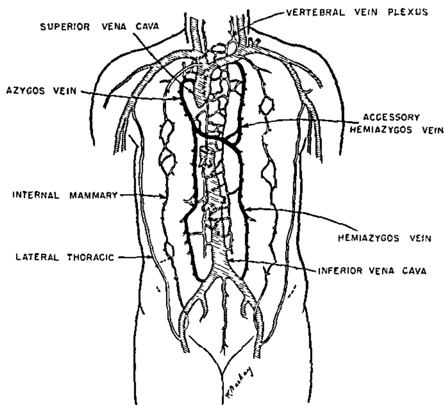


Fig 283 Schematic diagram of the venous circulation collateral to the vena cava system



Fig 284 Dilated hemiazygos system in a patient with portal hypertension The dilated veins are visualized behind the heart shadow From Leigh *et al* *Radiology* 63 696 1954

incident to cardiac decompensation Shuford and Weens described a case in which a hugely dilated azygos arch formed a distinct superior mediastinal mass measuring as much as 3 cm in diameter The engorged azygos vein shrank repeatedly with the relief of congestive heart failure

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Fig 285 Mediastinal varices in a 29 year old female with an 8 year history of splenomegaly The veins are visible as bilateral lobulated masses in the mediastinum Splenoportography reveals complete obstruction of the splenic vein with backflow of the contrast medium through a single greatly dilated and tortuous short gastric vein into the esophageal vein and azygos system From Molnar *et al* Splenic vein thrombosis *Radiology* 70 684 1958

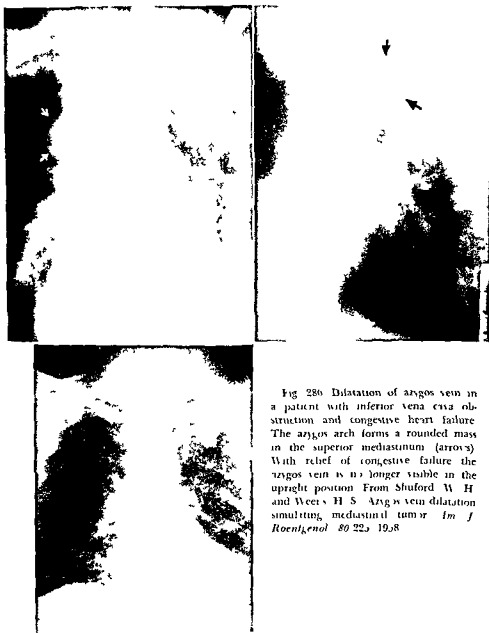


Fig 286 Dilatation of azygos vein in a patient with inferior vena cava obstruction and congestive heart failure The azygos arch forms a rounded mass in the superior mediastinum (arrows) With relief of congestive failure the azygos vein is no longer visible in the upright position From Shuford W H and Weens H S Azygos vein dilatation simulating mediastinal tumor *Am J Roentgenol* 80 225 1958

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Chapter 29

EXTRAMEDULLARY HEMATOPOIESIS

EXTRAMEDULLARY hematopoiesis is a condition in which blood is formed in areas outside the bone marrow (hetero topia of the bone marrow myeloid meta plasia) This condition is normal in em bryonic life but is presumed to be ab normal following birth Tumors of blood forming tissue may occur in adult life and at times these are seen in the mediastinum At least two other terms have been used in describing these tumors of hematopoietic tissue—hemocytoblastoma and erythroblas toma

In general the microscopic picture in extramedullary hematopoiesis resembles normal or hyperplastic bone marrow im mature forms of both the red and white series of blood cells as well as megakaryo *cytes are scattered through a mesh work* of fatty tissue

In fetal life hematopoiesis is normally carried on in many organs and tissues in cluding the liver spleen pancreas thymus lymph nodes adrenals kidney and thyroid At birth or soon thereafter however this extramedullary activity generally ceases and the marrow takes over the entire re sponsibility for blood formation Under the stimulus of excessive demand for blood cell production these extramedullary sites may resume their embryonic capacity Brannan points out that extramedullary hematopoiesis is fairly common in certain anemias of infancy and childhood and large tumor like growths of hematopoietic tissue may occur particularly in the biluses of the kidneys he states that this is to be regarded as a compensatory reaction (comparable to compensatory hypertrophy of a kidney)

In adults also this condition exists at times Blaisdell states the occurrence of extramedullary blood formation is to be regarded as a purely compensatory mecha nism in many cases in which as the result of destruction dysfunction or excessive re quirements the bone marrow is unable to meet the demands imposed on it It is found in association with many diseases among them widespread bone disease (par ticularly myelosclerosis and cancer) ane mias lead poisoning chronic nephritis hemochromotosis and various infections Most of the time the lesions are small and widely scattered in such areas as the retro peritoneal space liver spleen lymph nodes kidney pleura falx cerebri and others Brannan observes that the special sites of predilection in the adult for extramedul lary blood production have been very much *the same as in the young*

Occasionally one or more relatively large collections of extramedullary hematopoi etic tissues are seen in the mediastinum Saleeby reported a case wherein normal marrow was found in bilateral symmetrical nodules which were adherent to the fifth ribs near their vertebral attachments each of these masses measured 2 5 cm in di ameter and although attached to the ribs had no connection with the marrow cavi ties these lesions had a characteristic red dish color and resembled a blood clot Warren reported a case having a wide spread tumor extending through the retro peritoneal space mediastinum and else where and involving many lymph nodes Covey reported the case of an infant who died and who at postmortem examination had a 6 cm mass in the mediastinum which was attached to the pleura and intimatel connected with the hilum of the left lung histologically this tumor pure erythroblastic activity



Fig 287 Extramedullary *hematopoiesis in a 45 year old Spanish American male with an* acquired hemolytic anemia cause unknown Examination of the chest in 4 views (A B C and D) reveals lobulated masses in the lower mediastinum Thoracotomy disclosed that these lesions were attached to the pleura Histologic examination revealed a typical picture of extramedullary hematopoiesis Courtesy of Raymond R Lanier M D Denver Colorado

sonal communication with Brannan de scribed a kidney sized mass of bone mar row tissue in the thorax of a man who died of hemolytic jaundice (hereditary sphero cytosis)

Ask Upmark reported a case in which there were large bilateral paravertebral masses at the mid dorsal level which lay under the parietal pleura in the mediasti num against the vertebral bodies trans verse processes and the adjacent ribs these reddish masses of hematopoietic tissue had no connection with the marrow of the vertebra clinical and radiologic findings made the diagnosis possible in this case four years prior to postmortem examina tion Dawson reported some nodular growths in the paravertebral region of the mediastinum in a woman who had long standing acholuric jaundice with severe anemia Hartfall and Stewart s patient with acholuric jaundice and anemia had bi lateral reniform masses in the paraverte bral regions—on the right these extended from the seventh to the ninth dorsal ver tebra and on the left opposite the eighth and ninth these had no connection with the marrow of the vertebral bodies

Ask Upmark speculating on the possible origins of these masses in the mediastinum suggested the following (1) The matrix may be derived from patches of hematopoi etic tissue in the paravertebral regions (2) It may be derived from similar tissue in the pleura (3) It may be from lymph nodes (4) It may be derived from branches of the intercostal nerves

In adult cases of extramedullary hemato poiesis anemia has been a predominant clinical finding as it has been in infants

Radiologically it would seem that the findings in this entity are fairly character istic if they can be based on the gross pathologic descriptions of all reported cases of tumor like masses of extramedul lary hematopoietic tissue in the mediasti num and on the radiologic findings in at least two of these One would expect to see unilateral or bilateral masses of vari able sizes with smooth or lobulated bor ders lying in the posterior mediastinum against the vertebral bodies and adjacent ribs and confined within the limits of the parietal pleura but bulging it laterally These masses may be at any level There are no reports of calcium deposits within them

Extramedullary hematopoiesis should certainly be considered in any differential diagnosis when the radiologic picture fits the above descriptions particularly if there is jaundice or anemia

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Chapter 30

MISCELLANEOUS LESIONS

THERE IS A miscellaneous group of entities that occur very infrequently in the mediastinum Each of these deserves mention but are not of sufficient importance to warrant separate chapters They include plasmacytomas amyloidosis glomus tumors echinococcus (parasitic) cysts pseudocysts of the pancreas myxomas and xanthomas A short discussion follows on each of these

PLASMACYTOMAS

Plasmacytomas (plasma cell tumors) are solitary lesions composed of large numbers of plasma cells arranged in palisades and supported by a reticular stroma of connective tissue Most of them are found within bone but occasionally they occur as soft tissue tumors The extra osseous ones are seen most frequently in the upper air passages and oral cavity but the occasional case has been described in the mediastinum stomach intestine thyroid gland kidney ovary spermatic cord and skin (Childress and Adie)

Grossly plasmacytomas are soft to moderately firm in consistency and vary from *grayish white to grayish red in color* Stout quoting Oppikofer listed the following six categories into which true plasma cell *tumors might be divided (1) plasma cell* granuloma (2) benign localized plasmacytoma (3) malignant solitary tumor (4) malignant tumor *with generalization in* metastases (5) plasma cell myeloma and (6) plasma cell leukemia He pointed out



Fig 288 Plasmacytoma in a 37 year old male Examination of the chest discloses a mass in the posterior mediastinum immediately behind the left main stem bronchus Surgical exploration *revealed a 7 cm firm rounded* tumor which filled the space formed by the arch of the aorta The mass was removed but recurred 9 years later From Childress W G and Adie C C Plasma-cell tumors of the mediastinum and lung *J Thoracic Surg 19* 794 1950 and Childress W G and Adie G C Recurrent plasmacytoma of the mediastinum *J Thoracic Surg* 29 *480* 1955

the difficulty of recognizing which one of the six categories an individual lesion might be in when it first appears

Regional lymph nodes often become involved secondarily In a study of 104 cases of plasma cell tumors of the upper air passages and oral cavity Stout found no metastases in 54 cervical nodes in 24 and other nodes in 4 It is not unlikely that the mediastinal nodes are more often involved than generally recognized

The relationship between soft tissue plasma cell tumors and plasma cell myeloma is not clearly understood Stout concludes that it is impossible to know whether the bone lesions are manifestations of multiple primary sites of involvement or whether they represent true metastases from the soft tissue lesions

Radiologically, the few reported plasma cell tumors in the mediastinum have had no characteristic findings Those that have been described have been well circumscribed masses with smooth or lobulated borders All or most of these appear to be primary tumors in the mediastinum and not metastatic lesions in lymph nodes from some other primary site

At least one of the reported cases had an associated destructive lesion in a femur which was diagnosed as a localized myeloma On this basis it is suggested that when myeloma is diagnosed a chest examination always be made for the possibility of mediastinal tumefactions

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AMYLOIDOSIS

Amyloidosis is a disease of unknown etiology in which there is deposited in tissues a waxlike homogeneous material rarely such masses are found in the mediastinum It is probably a degenerative process but in some instances there is evidence that it may be infiltrative

Schmidt *et al* have classified amyloid disease as follows

1 Primary amyloidosis a rare form in which there is no obvious pre existing disease this type has 2 forms—systemic and tumor forming

2 Secondary amyloidosis accompanying such entities as infections (tuberculosis empyema bronchiectasis pulmonary abscess and others) tumors (carcinoma lymphoma leukemia and in particular multiple myeloma) and miscellaneous conditions such as rheumatoid arthritis diabetes thermal burns and cirrhosis of the liver

Primary amyloidosis involves chiefly the tissues of mesodermal origin Localized infiltrations are prone to occur in the heart skin larynx trachea mediastinum and bronchi (Schmidt *et al*) Cardiac involvement is the cause of death in one half of the cases (Dahlin) Secondary amyloidosis involves chiefly the liver spleen kidneys and adrenals Both the primary and secondary types occasionally involve the lymph nodes Mediastinal involvement may result either from this lymph node invasion or through infiltration of the connective tissues

Radiologically when the mediastinum is involved in amyloidosis there may be generalized widening resulting from diffuse involvement of the connective tissues or of the lymph nodes or there may be tumefaction resulting from a localized involvement

Schmidt *et al* have reported one proven case in which the localized mediastinal amyloid mass contained calcium and another unverified case in which the mass compressed the lower esophagus Mediastinal involvement is usually accompanied by

ulmonary involvement which may appear s diffuse linear infiltrates pneumonic con olidation involving one or more lobes r hilar adenopathy

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GLOMUS TUMORS

Glomus bodies are structures normally ound in the body and are principally lo ated in the extremities in such areas as he palmar and plantar aspects of the hands nd feet and beneath the nails of the fin ers and toes they are found to a lesser xtent in other parts of the body Occa ionally as a result of hyperplasia of their lements they enlarge to become tumors Although this usually happens in the ex tremities at least 1 case has been described which arose in the posterior mediastinum (Brindley)

The normal glomus consists of an af ferent arteriole an anastamotic vessel col lecting veins non medullated nerves and a reticular supporting structure These bod ies are thought to aid in the regulation of heat and in the control of blood pressure In about half the cases where hyperplasia has developed trauma has seemed to play a part

Brindley s patient was a 29 year old fe male whose chief complaint was that of pain in the right lower chest of 9 years duration The past history revealed that a tumor of 1 cm diameter and purple in color had been removed from the right groin 2 years earlier There were 4 similar masses in the right lower extremity these were rounded elevated tender and purple in color Laboratory data were negative Radiologic examination of the chest re vealed a mass in the left posterior mediasti num at the level of the eighth ninth and tenth dorsal vertebrae Surgical exploration



Fig 289 Glomus tumor of the mediastinum The case is described in the text The chest xamination discloses a posterior mediastinal mass (not visible here on the reproduced frontal iew) lying to the left of the eight ninth and tenth dorsal vertebrae From Brindley G V ,lomus tumor of the mediastinum J Thoracic Surg 18 417 1949

disclosed a 5 cm lesion attached to the eighth intercostal nerve Shortly after the removal of this tumor the 4 tumors of the right lower extremity were also removed Microscopic examination of the mediastinal mass and of those removed from the right leg were identical and were reported as glomus tumors by the pathologist

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ECHINOCOCCUS CYSTS

Echinococcus (parasitic) cysts occur very rarely in the mediastinum although they are relatively common in the lungs and pleural space of patients afflicted with hydatid disease Heuer and Andrus cite 8 cases where the mediastinum was involved Seven of these had hour glass lesions involving the spinal canal and adjacent para vertebral space The other also had an hour glass lesion consisting of a large cyst in the anterior mediastinum which connected by a stalk to a smaller cyst in the adjacent anterior thoracic wall

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PSEUDOCYST OF THE PANCREAS

Edlin has reported a case of pseudocyst of the pancreas which extended upward from the abdomen into the mediastinum through the esophageal hiatus The pseudocyst was associated with chronic inflammation and multiple intraabdominal pseudocysts of the pancreas

The patient was a 60 year old male admitted to the hospital with clinical findings of heart failure long standing chronic pancreatitis and diabetes Radiologic studies disclosed a large mass in the lower mediastinum behind the heart which broadened the mediastinal borders both to the left



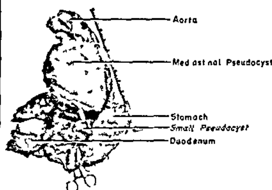


Fig 290 Pseudocyst of the pancreas projecting into the mediastinum The case is described in the text A heavily penetrated chest roentgenogram discloses a fusiform shaped retrocardiac mass in the mediastinum From Edlin P Mediastinal p of the pancreas *Gastroenterology* *17* 96 1951

and to the right there was also a moderate amount of fluid in the left pleural space and left ventricular hypertrophy

The patient died during his hospital admission and necroscopy disclosed a 15 by 30 by 10 cm cystic mass in the posterior mediastinum lying between the parietal pericardium and the esophagus and aorta Superiorly the mass extended to the tracheal bifurcation and was attached to the left main stem bronchus Inferiorly it extended into the abdominal cavity through the esophageal hiatus passing through the lesser peritoneal sac to the superior border of the pancreas The cyst wall measured 0 5 cm in thickness and its lining was smooth and glistening The pancreas had many other abdominal cysts associated with it

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MYXOMAS

Myxomas are tumors of the fibroblast *showing a marked secretion of mucoid material* Those of mediastinal origin which have been reported have been large in size and variable in position They tend to be *well encapsulated One of* those in the anterior mediastinum was dumbbell in type *with a soft tissue component presenting in* the anterior chest wall as a small palpable mass near the right nipple

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XANTHOMAS

Xanthoma is a confusing term since it is used to describe both neoplastic and non neoplastic processes The non neoplastic lesions are accumulations of lipoid laden macrophages whereas the truly neoplastic xanthomas represent changes in fibromas or other connective tissue tumors

The few reported cases of neoplastic xanthomas in the mediastinum have been well encapsulated lesions of variable size arising in the costovertebral groove posteriorly On radiologic examination they tend to resemble encapsulated neurogenic lesions with the exception that there is very little if any tendency for them to erode or destroy bone or to encroach upon the spinal canal

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